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THE DISEASES OF CHILDREN

A WORK FOR THE PRACTISING PHYSICIAN

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THE DISEASES OF CHILDREN

DISEASES OF THE UROGENITAL SYSTEM

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DEVELOPMENT OF THE GENITO-URINARY SYSTEM, DEFECTS OF FORMATION, HERMAPHRODITISM

THE fact that organs of such different physiological importance as are represented by the suprarenal glands, the urinary apparatus, and the sexual organs, form one evolutionary unity is shown, not only by their pathology, but by their topographical relations. It seems necessary, therefore, to discuss in one common section the diseases of these three systems.

FIG. 1.

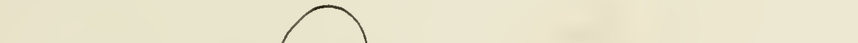


Diagram showing the indifferent disposition of the genito-urinary system of a mammal at an early stage. (According to O. Hertwig.)

The disorders of development (defects of formation, anomalies) have not only a theoretical interest, to which they are eminently entitled because their study helps our understanding of the mechanism of evolution, but the practical physician, too, should not undervalue their importance. This will be shown—among other things—by the discussion of hermaphroditism, and of those deformities whose early diagnosis provides a life-saving therapy.

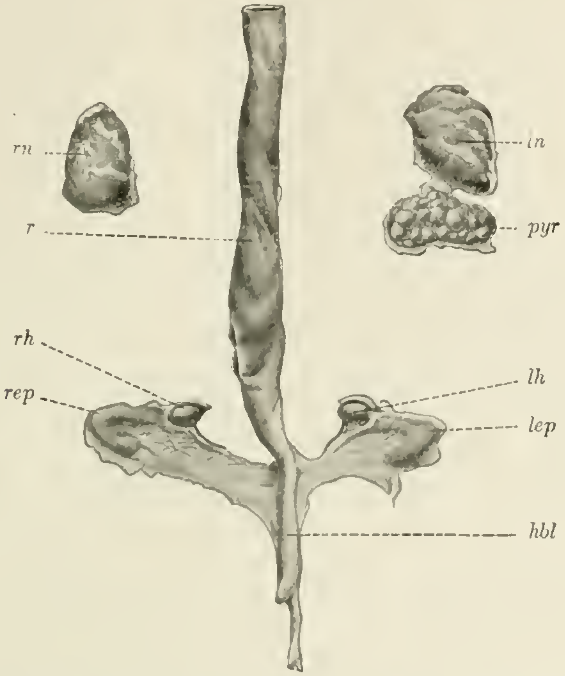
It is probable that the greatest number of congenital deformities result from arrest of development; their mode of origin can therefore be understood only by a knowledge of the normal process of development. This will be illustrated by the following table taken from Neugebauer, the comprehension of which will facilitate the understanding of the sketch drawn by Hertwig.

COMPARATIVE EMBRYOLOGICAL TABLE.

Male genitals.	Urogenital organs of fœtus at indifferent stage.	Female genitals.
Testicle, ampullæ seminales, canaliculi seminiferi.	Germ epithelium (genital gland).	Ovary: Graffian follicle.
	Pronephros (Oken's, Wolff's body = mesonephros).	
(*a*) Epydidymis, rete testiculi, tubuli recti, vasa efferentia, testis, hydatis pedunculata epididymidis.	(*a*) Upper part — pars sexualis.	(*a*) Epoophoron (parovarium, Rosemüller's organ), medullary sheaths of the ovary, rete ovarii and hydatis pedunculata ovarii.
(*b*) Paradidymis (Giraldés organ, vasculum aberrans Halleri).	(*b*) Lower part — metanephrotia.	(*b*) Paroophoron.
Cauda epididymidis on either side, vasa deferentia, vesiculæ seminales, ductus ejaculatorii.	Wolffian ducts (emptying ducts of the pronephros).	Malpighi-Gartner's canals, going centripetally into the ligamentum lata, near the internal os of the uterus, or entering somewhat above into the inner walls of the womb, running in these to the vaginal parts of the uterus, where some side retroussments form the analogues to the vesiculæ seminales; then going, convergent, in the lateral and anterior partition walls, into the urethral folds. (Both canals normally disappear, but when persistent lead to the formation of cysts.)
Hydatis sessilis testiculi on either side, sinus prostaticus in capite gallinaginis (utriculus masculinus — vesicula prostatica).	Müllerian fibres.	Tubes with fimbriæ and hydatis Morgagnii on either side, uterus, vagina.
Kidneys and ureters............	Metanephros (permanent kidney) and ureters.	Kidneys and ureters.
Gubernacula Hunterii..............	Pronephros—inguinal ligaments.	Lig. ovar. propria, lig. rotunda uteri.
Pars membranacea urethræ	Sinus urogenitalis.	Vestibulum vaginæ.
Penis.....	Genital tubercle.	Clitoris.
Halves of the scrotum	Genital folds.	Labia pudenda majora.
The lateral walls of the penile urethra and of the corpus cavernosum penis.	Genital furrows.	Labia pudenda minora and the so-called "bride masculine," "bride penienne," the bright tissue string running from the urethral orifice to the base of the clitoris.

The deformities involve either the whole complex of the genito-urinary apparatus, including the terminal portion of the bowel, or they are restricted to a single organic system or organ. The first are of rare occurrence and of little practical interest, as they are generally found in still-born children. The following cut gives an illustration of such a total deformity.

Fig. 2.



Deformity of the urogenital system of a newborn boy. Atresia ani vesicalis. Rectum (*r*) emptying into the string shaped bladder (*hbl*); *rh*, and *lh*, right and left testicle; *rep*, and *lep*, right and left accessory testicle ending sausage shaped. Prostate, ureters, seminal vesicles are lacking. *pyr*, polycystic rudiment of the left kidney, resulting from cystic degeneration of the tubuli contorti, which did not become connected with the absent tubuli recti. *rn*, right kidney. *ln*, left kidney.

Of greater practical importance are those defects of formation that are limited to a single organ or organic system. We recognize those of the kidney, of the urine-conducting tracts, of the male and female sexual organs, and of the terminal portion of the bowels. They will be described in speaking of the pathology of these organs. For the present it will be convenient to appreciate the theoretical and practical importance only of that deformity of the genital apparatus which consists in a persistence of the male and female organs in the same individual—the so-called HERMAPHRODITISM.

Pathology distinguishes between TRUE and FALSE HERMAPHRODITISM. The first name is applied when male and female genital glands exist side by side; the latter when the construction of the genital glands conforms to one type, while the genital ducts and the external genitals conform to the opposite type.

The *genesis of hermaphroditism* will be clearly understood if we remember that the fœtal disposition of the genital ducts is bi-sexual

(Waldeyer). Every embryo possesses Wolffian ducts and Müllerian fibres, and the development of the germ glands, indifferent during the first fœtal period, may progress in the same individual in two directions. The existence of true hermaphroditism, which was for a long time denied, is proven by the famous observations of v. Salen and Garré. They concern individuals in whom typical ovarian and typical testicular tissue were found in the same genital gland. Of course both varieties of tissue were undeveloped and were not fit for functional action, and true hermaphroditism can therefore claim only theoretical interest, since—setting aside its great rarity—up to this time the faculty to impregnate and to conceive simultaneously has never been observed in man. The common classification into hermaphrodismus lateralis, unilateralis and bilateralis, does not require any comment.

Pseudohermaphroditism is far oftener found. Neugebauer who may be considered to be the best authority on this subject, has analysed 988 single observations, and reported 43 cases in his own practice. He recommends that we preserve to-day the old division of Klebs into masculine and feminine hermaphroditism. Either of these may be internal, external, or complete. Neugebauer characterizes these variations as follows:

I. PSEUDOHERMAPHRODISMUS FEMININUS

A. Internal.—External genitals are normally feminine in development. Beside the ovaries and more or less perfectly developed genital ducts, Wolffian ducts may also be more or less developed (rarest form of pseudohermaphroditism).

B. External.—The clitoris resembles a penis; the more or less deformed labia pudendi appear like the empty scrotum, and in one or both halves of this seeming scrotum rounded bodies are sometimes palpable, and it is therefore easy to make a mistake in determining the sex. These formations may be either ectopic ovaries, a hydrocele, an hæmatocele, or even a hernial protrusion. This form of female pseudohermaphroditism sometimes imitates a peni-scrotal hypospadias.

C. Complete.—External genitals more like the masculine. Besides the ovaries, both Müllerian fibres and Wolffian ducts are more or less developed (rare form).

II. PSEUDOHERMAPHRODISMUS MASCULINUS

A. Internal (more frequent than the feminine form). External genitals formed normally masculine. In the pelvis, beside normally or rudimentary formed Wolffian ducts and their derivatives, Müllerian fibres are also found more or less developed: uterus with tubes, ligaments, and vagina. The latter opens either into the prostatic part of the urethra, or between the halves of the scrotum, in which latter case the urethra empties into the vagina. If a uterus is present, the testicles are very frequently located where the ovaries would lie in woman. The unilateral or bilateral chryptorchism stands in a casual relation to the presence of a uterus in man.

B. External (the most frequent form of pseudohermaphroditism). The vulva is imitated in consequence of peni-serotal hypospadias, with more or less rudimentary development of the hypospadiac penis. There are many cases where upon such a vulva, not the least variation from the normal vulva hints at a possible "erreur de sexe," in case the individual should have been christened a girl. In some cases there is hypospadias of the penis only, or the scrotum is only partly split, and the orifice of the urethra lies in its upper half, in other instances the scrotum is completely divided, and even little rudimentary labia pudendi are imitated by the split penile urethra. In most cases the false clitoris seems to be enlarged, but there are examples where the clitoris seems small even for a woman, and is therefore quite rudimentary in appearance. The internal genitals are masculine in formation, analogous to the testicles. The cases are very rare in which the penis is normal, and the scrotum alone shows a split in its lower part, where the opening of a vagina appears to be located.

C. Complete.—Besides the seemingly feminine external genitals there are to be found more or less developed Müllerian fibres, together with the presence beside the testicles of more or less developed Wolffian ducts: *i.e:* Uterus with tubes and ligaments, a vagina which often empties into the urethra, or in common with the urethra into the sinus urogenitalis, which is persistent, or else the urethra empties into the vagina.

In the six recorded cases where a juxtaposition of hetero-sexual external genitals occurred: *i.e.* where scrotum and penis existed along with a vulva, there probably was, in the opinion of Neugebauer a split in the lower end of the vertebral column (dipygia). Hermaphroditism is of great theoretical interest in embryology, and in consideration of the many questions of civil and criminal law which may arise in connection with the life and destiny of hermaphrodites. But setting aside all this, it was necessary for the present purpose to state in detail Neugebauer's observations because the children's physician may often be called upon to answer the query of parents as to the sex of a newborn child. (Three times in the last 4000 cases at the Berlin University Polyclinic for Children a decision was possible, if at all, only when the relations just discussed were fully understood).

If we remember that in masculine hermaphrodites the feminine appearance is simply imitated by peni-scrotal hypospadias (see chapter on the Male Genitals), it will be clear that the masculine sex can be established only by the presence of testicles, accessory testicles, and spermatic ducts. It is out of the question to consider the presence of the prostate, setting aside the difficulty of recognizing it by palpation, as a very important element in differential diagnosis. "The masculine sex is assured if it be possible to touch in the imitated labia pudendi symmetrical formations corresponding in size, shape, and consistency to testicles, and accessory testicles, even though the opening of a vagina, surrounded by hymen, should be found beneath the imitated feminine mouth of the urethra, and even labia pudendi minora as indications of the edges of the split penile urethra." Neugebauer from his experience does not approve of the dictum of Klebs that the presence of labia pudendi minora is decisive for the feminine gender. He says, "If in the newborn it be not possible to feel assured of having touched the testicles, and accessory testicles. the only proper thing to do is to declare until later that the sex is doubtful, to demand a future repetition of the examination, and finally even to postpone the decision until the individual has arrived at the age of puberty. But if the case is one with cryptorchism on both sides the opinion must certainly be postponed or else a serious error will be risked."

The physician will the more readily decline to give a diagnosis which influences decisively the education of the patient, when he recalls those cases in which a testicle of the masculine pseudohermaphrodite was erroneously considered to be an ectopic ovary of a female, or those cases in the formations which during life were believed to be testicles were found at autopsy to be something entirely different, *e.g.*, the case found by Virchow to be a pre-inguinal hernia of the processus vaginalis peritonei. The laws of every state should recognize the fact that it is sometimes utterly impossible to determine the sex exactly, and should make allowance for doubtful sex (Unger).

Atresia ani and atresia urethræ are not rare among the deformities which complicate hermaphroditism, and it is important for the physician to be aware of this, since they require an immediate surgical operation.

DISEASES OF THE SUPRARENAL GLANDS

EVOLUTION, ANATOMY, AND PHYSIOLOGY

Our knowledge of the evolution of the suprarenal glands is still deficient. Although the opinion prevails that the cortical substance proceeds from the pronephros, while the medullary substance is genetically connected with the sympathetic nervous system, there are still authors who deny any evolutionary connection between the pronephros and the genito-urinary system. They support their opinion by the fact that among other things, the suprarenal glands may be preserved even if the urinary apparatus is completely lacking, and that, these organs do not participate in the congenital change of situation of the kidney. There is a time in life when the suprarenal gland is larger than the kidney. In adults the relation of the suprarenal to the kidney is as 1: 28; in the newborn it is as 1: 3.

A section through the suprarenal gland shows the division into two portions, which are distinguished by their color: the yellowish and radially striped being the cortical substance, and the grayish red and spongy being the medullary substance. The latter consists of cells which will turn intensely yellow or brown when treated with chromic acid (chromatin cells), as shown by Kohn, of Prague,—a fact of the greatest importance since it was instrumental in showing the wide extension of these cells, which compose the medullary substance, throughout the human organism, and especially of the newborn. The latter—as shown by the examinations of Kohn, Zuckerkandl, and Wiesel—is extraordinarily rich in chromatin tissue, which has evidently the same physiological significance as the medullary substance of the suprarenal glands. Zuckerkandl found, beside the inferior mesenteric artery, in the embryo and in the newborn, an oblong, oval formation of which the most essential elements are chromatin cells (by-bodies of the sympathetic). Its structure corresponds to the medulla of the suprarenal glands, but it does not contain ganglionic cells. Wiesel discovered a surprising abundance of chromatin tissue in the retroperitoneal space of the newborn, in the cœliac plexus, and near the exit of the inferior mesenteric artery. (Part of this tissue will later retrograde to the sympathetic.) The testicles of about 75.5 per cent. of the newborn contain, according to Wiesel, chromatin tissue.

The fact that the whole mass of the chromatin system represents a complex of cells of which the function is a unit, promises to become of importance in pathology, because physiological and chemical investigation have shown that it is only from the medullary portion of the suprarenals, and from other chromatin tissues, that there can be obtained the substance which, when introduced into the animal body (even in minute quantity, fraction of a milligram), can raise the blood pressure enormously. This is the so-called adrenalin which was crystalized out by Takamine, and it seems as if we shall soon ascertain its constitution and synthesis. This body cannot be developed from the cortex of the suprarenal gland, which is of greater importance to life than the medullary substance, as has been shown by experiment. The importance of the cortical substance in the normal processes of life is said to be its antitoxic action, but this is still hypothetical.

APLASIA AND HYPOPLASIA OF THE SUPRARENAL GLANDS

Congenital absence of the suprarenal appears to be exceedingly rare. When it is accidentally discovered at autopsy on an otherwise normal person, we suspect that accessory suprarenal glands, which have vicariously undertaken the function, have been overlooked. (Marchand's suprarenal glands,—*i.e.*, the suprarenal tissue which is found near the spermatic veins, and normally, in the male along the spermatic ducts, in the female, in the lateral ligaments.) According to Schmorl, accessory suprarenal glands are found in 92 per cent. of human beings.

Hypoplasia of the suprarenals is often associated with deformities of the central nervous system (hemicephalia, encephalocele, etc.).

Zander is of the opinion that the anterior portion of the cerebrum exerts a trophic influence over the development of the suprarenals. Ilberg, who described a case of aplasia of the suprarenals, combined with hemicephalia, propounds the following hypothetical explanation for the frequent association of cerebral deformities with aplasia of the suprarenals:—that the hæmadynamic function of the suprarenals has been lacking through underdevelopment, and that in consequence the brain, which was not protected against anæmia, developed badly.

HYPERÆMIA AND HÆMORRHAGE OF THE SUPRARENAL GLANDS

In the newborn, the suprarenal blood supply is normally very rich. This fact explains its very pronounced tendency to passive hyperæmia in all those diseases which are attended by general congestion; in all infectious diseases hyperæmia is very active. It has been long recognized that the experimental infection of guinea-pigs with diphtheria would result in hyperæmia of the suprarenals, with hæmorrhagic infarction, a condition which we sometimes find also at the necropsy of children who have died of diphtheria. Hæmorrhages of the suprarenal glands (suprarenal apoplexy) are not rare in the newborn. They may be unilateral or bilateral. The suprarenal gland often attains the size of a hen's egg, and its parenchyma often appears to be completely destroyed. These conditions are shown in the colored illustrations of Plates 56 and 57.

In this case, that of a newborn boy, both suprarenal glands were changed into hæmatomata of the size of a pigeon's egg. The autopsy showed also a fracture of the left humerus, a separation of the cartilages from the second to the fifth ribs and a hæmorrhagic infiltration of the left sternomastoid, and also of the neighboring fatty tissue.

The conditions in this case illustrate one of the causes of apoplexy of the suprarenal glands, *i.e.*, trauma resulting from obstetric manipulations in difficult delivery, or in reviving asphyxiated children. But still another series of circumstances may be responsible for the formation of hæmatoma of the suprarenal glands some of which may affect, not only the newborn, but older children as well. Such are compression of the inferior vena cava between the liver and the vertebral column, caused by strong contraction of the uterus, by which the infantile abdomen is influenced; compression of the umbilical cord during delivery; acute degeneration of the vessel walls; degeneration of the tissue of the suprarenal glands; convulsions; syphilis; vasomotor disturbances in cerebral diseases; thrombosis of the renal veins and the inferior vena cava; infections. Hamill is of the opinion that hæmorrhage of the suprarenal glands occurring before birth is generally caused by the act of delivery; and that those hæmorrhages which occur post-partum are as a rule the result of infection spreading from the remains of the umbilical cord.

The etiology of some of the reported cases, especially of those occurring in older children is very obscure. The case of a child who suffered from purpura hæmorrhagica, and died of suprarenal hæmorrhage seems to be of decided interest.

The diagnosis can seldom be established except at the autopsy, since the children generally die of collapse; less frequently after several days, with symptoms resembling peritonitis. Addison's complex of symptoms is not often observed, evidently because of the brief duration of the disease. Dissections often show the presence of free hæmorrhage into the abdominal cavity.

TUMORS OF THE SUPRARENAL GLANDS

Steffen has collected nine cases of carcinoma and sarcoma of the suprarenal glands in infancy, in which cases he found them in the primary stage. According to his statistics even the earliest infancy may develop malignant tumors of the suprarenals.

Otto Ramsay mentions twenty-five cases of carcinoma of the suprarenals, of which five occurred between one and twenty years, and twenty-six cases of sarcoma, of which eight developed between six months and twenty years.

The growth may be either acquired or congenital. Tumors of the suprarenals are extraordinarily disposed to hæmorrhage and to metastasis. Death occurs as the result of profound cachexia. Addison's complex of symptoms may be present, and may establish the correct diagnosis. Linser has related the history of a five-year-old boy, whose bodily development and sexual organs resembled those of a youth of sixteen and eighteen, but who had only learned to speak six months before. He was brought to the clinic on account of a malignant adenoma of the left suprarenal gland, which had extended to the renal vein and vena cava.

Upon the basis of this case, Lisner seeks the cause of giant growth in excessive function of the suprarenal glands.

Without laparotomy, it is hardly possible to differentiate tumors of the suprarenal glands from other abdominal tumors. (For the differential diagnosis of suprarenal tumors see special section.)

ADDISON'S DISEASE

This disease has been brought by its discoverer into casual relation with changes in the suprarenals. In its essential parts his original description is probably still perfectly valid. Neusser, to-day probably the best authority on the subject, characterizes the disease as follows:—"There is idiopathic anæmia, accompanied by great adynamia and apathy, disturbances on the part of the digestive tract and the nervous system, and accompanied by a bronze discoloration of the skin. The disease has a chronic course, with progressive cachexia, but often shows turbulent symptoms, such as intractible diarrhœa, coma, or convulsions, and terminates inevitably in death."

The disease is rare in infancy. Monti has found among 290 cases, 11 in children. Family tendency has not been established.

Symptomatology and Diagnosis.—In children there is emaciation, pallor, and asthenia, but the asthenia is greater than would be expected from the degree of wasting. Gastro-intestinal disturbances gradually appear which may vary from the mildest indigestion to the most intractible vomiting and profuse diarrhœa. Pigmentation, which in most cases determines the diagnosis, often appears quite late. The pigmentation varies widely in degree. Sometimes there are only a few pigmented spots upon the surface of the palate, which may be only too easily overlooked, so that, wherever Addison's Disease is suspected this region should receive the closest scrutiny. In other cases the pigmentation is so pronounced that almost the entire cutaneous surface is bronze-colored.

The temperature may be subnormal, but febrile paroxysms have also been described. Nervous symptoms, headache, vertigo, convulsions,—are part of the clinical history. According to Gerhardt, convulsions are seen in two-fifths of the cases in childhood. Acute paralyses (peroneus palsy) also occur.

The duration of the disease is variable. Generally it lasts months, less frequently years. In one case, described by Netter, the process was very acute. The child was four years old, perfectly healthy, and was suddenly taken ill with vomiting, diarrhœa, apathy, and depression. A diagnosis of peritonitis was made, and the child died after three days, when the autopsy revealed tuberculous foci in the suprarenal glands. This case illustrates the great difficulty of establishing the exact diagnosis in diseases of the suprarenals, for the very reason that their symptoms are so extraordinarily ambiguous.

Leube, in his well-known "diagnosis" remarks in regard to the suprarenals that their diseases are not as yet subjects for diagnosis.

Pathological Anatomy and Pathogenesis.—In almost all cases of Addison's Disease the suprarenals have been found to be affected by tuberculosis. The tuberculous infection was either total in both glands so that there was no normal tissue to be found, or else small foci existed in one or both organs.

Tuberculosis of the suprarenal glands is distinguished by abundant formation of granulation and scar tissue. The process may be eminently chronic, until finally the entire suprarenal gland may be changed into scar tissue, in which only sparse caseous portions may still be discovered, or the scar formation may extend through the capsule of the suprarenal, and affect the cœliac axis as well, a fact which may have an important bearing upon pathogenesis.

Other pathological processes of the suprarenals (tumors, atrophy, etc.) were found in a small number of cases.

The fact that there do undoubtedly occur typical cases of this disease in which the suprarenal glands are sound, made it necessary to seek the cause of Addison's Disease not only in disturbance in the functions of the suprarenals, but also in those of the sympathetic nervous system. From a careful analysis of heretofore observed cases, and on the ground of the results obtained by experimental pathological research, and by physiological chemistry, Neusser arrives at the following opinion as to the character of Addison's Disease:—"The suprarenal gland is one which produces an internal secretion, whose function is to counteract the toxic products of the metabolic activity of other organs, and to produce a substance which is indispensable above all to the preservation of the normal tone and to the nourishment of the sympathetic system. Addison's complex of symptoms is in every case dependent upon injury to, and finally entire suspension of the function of the suprarenal glands. This may result either from an anatomical disease of the glands themselves, or else their secreting and antitoxic action may be hindered and finally paralysed by disturbance of the conducting tract which controls their function. This tract runs from the spinal marrow through the splanchnic and cœliac ganglion. In this manner the result will be on the one hand a nutritive and functional disturbance of the sympathetic system, and on the other, a general auto-intoxication. Besides these two principal factors, a local damage to the abdominal sympathetic, by extension of the pathological process to it, plays in many cases a part in the production of sundry Addisonian symptoms. Pigmentation of the skin and mucous membranes is not an integral part of Addison's complex; it may have diagnostic, but it does not have absolute importance. Pigmentation is not a direct, but an indirect symptom of suprarenal disease. It is caused only through the medium of the sympathetic, damaged generally or locally."

As to the **pathogenesis** of Addison's Disease, even at this day, opinions are widely at variance. Wiesel from the examination of five cases, sees the essential factor in the complete destruction of the cells of the chromatin system. Karakascheff, who had worked out the problem in Marchand's Institute, awards to the cortex of the suprarenals the whole importance in the origin of the disease. He infers this from the observation he made upon a five-year-old child who died of atrophy, independent of any disease of the suprarenal gland, whose autopsy revealed extensive hæmorrhage, which had existed since birth, in the medullary substance, which was thereby completely destroyed. The cortical substance was intact. According to this, it would seem that the lowering of blood pressure which can always be proven clinically, might explain the relation of the medullary substance to Addison's Disease.

Therapeutics.—Up to this time there is no record of recovery from this condition. Organotherapy with the numerous commercial preparations on the market has signally failed. The gastro-intestinal and nervous manifestations must be treated symptomatically.

FUNCTIONAL DISTURBANCES OF THE SUPRARENAL GLANDS IN GENERAL DISEASES, INFECTIONS AND INTOXICATIONS

Luksch has pointed out that the participation, in a series of diseases, of the suprarenal glands, as organs having an internal secretion, cannot be without influence upon the progress of these diseases. It is of interest to the pediatrist that Luksch, in agreement with the observations of earlier writers, has succeeded in proving that the diphtheria toxin affects the suprarenal glands so powerfully that they are completely or partially deprived of their capacity to raise the blood pressure. Luksch is of the opinion that many a death from diphtheria finds its explanation in this deprivation of the function of the suprarenals.

DISEASES OF THE URINARY APPARATUS

In no case should the examination of the urine be neglected. Even though the physician does not suspect from the history the existence of disorders of the urinary apparatus, yet he cannot fulfil his duty without a simple uranalysis. Emphasis may well be laid upon this fundamental law of medical examination at the very beginning of this chapter. The reason for this is, on the one hand that it is peculiar to a number of the affections of the kidneys in infancy that they produce symptoms of general disease, while their own existence might in no wise be suspected a priori, on the other hand, the difficulty of obtaining a specimen of urine from a young child, and especially from an infant, is very apt to induce the physician to neglect its examination.

The difficulty of obtaining specimens of urine from infants is probably the reason why our knowledge of its physiological and pathological chemistry consists only of scientific fragments.

THE PHYSIOLOGY AND PATHOLOGY OF THE URINE

Daily quantity.—During the first three or four days of life the secretion of urine is extremely small. This is not by any means solely due to the lack of nourishment, for babies suckled by a nurse and artificially fed infants, both of whom receive relatively plenty of liquid, behave exactly the same as those who are nursed at the mother's breast. The considerable individual variations in the urinary secretion during the first days of life, as shown in the comparative table of Reusing, are remarkable.

	Minimum	Maximum
In the first 24 hours after delivery	2 c.c.	61 c.c.
2nd day	11 c.c.	145 c.c.
3rd day	13.3 c.c.	171 c.c.
4th day	17.5 c.c.	179 c.c.
5th day	22.5 c.c.	222 c.c.
6th day	70 c.c.	280 c.c.
7th day	93 c.c.	338 c.c.
8th day	100 c.c.	331 c.c.

In comparing the quantity of the urine and the quantity of nutriment, and in computing the proportion of the percentage, we find that the sucking babe has no continuous increase in the daily quantity, but tolerably irregular variations. It seems pretty well established that the nursling during its first days of life secretes relatively little of the water introduced, while the quantity of the urine of the artificially fed babe attains both absolutely and relatively high quantities. This will be illustrated by the figures given by Reusing.

Day.	Quantity of milk.		Quantity of Urine		Urine: Percentage to quantity of milk.	
	Bottle-fed.	Breast-fed.	Bottle-fed.	Breast-fed.	Bottle-fed.	Breast-fed.
1	96.0	38.3	35.8	8.4	37.0	21.8
2	150.0	120.8	71.0	26.8	47.0	22.2
3	229.5	176.6	135.8	40.9	58.8	23.0
4	253.1	220.0	187.0	60.8	74.0	27.6
5	364.6	271.5	283.0	119.1	78.1	43.9
6	369.0	296.0	246.0	148.6	66.6	50.0
7	410.0	297.0	325.0	157.0	79.1	57.6
8	530.0	338.0	406.0	208.0	77.0	62.5

But even in the artificially nourished babe, the increase of urinary secretion does not exactly correspond to the increased supply of food.

Premature infants secrete minimal quantities of urine, but we do not have many subjects. In the later months of the first year, for the healthy babe the law becomes valid that 100 Gm. of nourishment produce 68 c.c. of urinary secretion. This law was discovered by Camerer, and confirmed by Czerny and Keller. In regard to the quantities of urine secreted by healthy infants after the first year, Camerer has fully informed us by exact examinations conducted through many years.

They show that the increase in the quantity of the urine is not exactly in proportion to the nutritive supply, but lags behind it. It is of course not possible to formulate a rule for the quantity of urine at every stage of life: a mixed diet, as well as individual peculiarities, will cause a variation in the proportions. The table prepared by Holt which takes cognizance of the examinations of Schabanowa, Cruse, Camerer, Pollack, Martin-Ruge, Berti, Schiff, and Herter, has therefore only a relative value. According to Holt the daily quantities of urine are:

In the first 24 hours up to	60
In the second 24 hours	10–90
From third to 6th day	90–250
From 7th day to 2 ms.	150–400
From 2 ms. to 6th month	210–500
From 6th month to 2nd year	250–600
From 2nd to 5th year	500–800
From 5th to 8th year	600–1200
From 8th to 14th year	1000–1500

Very different pathological conditions produce changes in the quantity of the urine. It must therefore be most emphatically stated that the observations of Camerer are valid only for the healthy child, and even slight disturbances may change the proportion between the quantity of urine and the quantity of nutriment. Granular atrophy (contracted kidney), diabetes mellitus and insipidus, which may begin even in the nursling, will result in the excretion of especially large quantities of urine. It may de very difficult to establish by uranalysis the difference between granular atrophy (contracted kidney) and diabetes insipidus. Hypersecretion and low specific gravity are characteristic of both diseases, and renal casts may be equally lacking in the contracted kidney during infancy.

Frequency of Urination.—In the newborn the bladder is emptied as a rule immediately after delivery (Bendix). The younger the child, the oftener the bladder is emptied, but it is possible by training to bring about an early control over the sphincter. According to Holt a child of two years can retain its urine for from 2–6 hours during sleep; a normal child of three years may sleep for 8 to 9 hours without wetting itself. and while awake may retain the urine for intervals of 2 to 3 hours.

Physical Properties.—The color of the urine of healthy infants is generally a pale straw-yellow, perhaps a little more intense up to the sixth to tenth day, but still lighter in color than the urine of adults. The concentration (or specific gravity) generally influences the depth of the color. The reaction of the urine is strongly acid during the first few days, especially, according to Flensburg, during the period of inward infarction. The urine of the newborn either does not normally turn the plane of polarized light to the left, or else does so quite feebly. According to Czerny and Keller, the specific gravity of the healthy infant varies according to the state of nutrition, from 1.011 to 1.012. "As soon as the supply of liquid and the excretion of the urine have reached the normal quantity, the specific gravity sinks to 1.003–1.004, and remains at this level till the end of the first year."

Holt gives the following summary of the specific gravity at the various periods of life:—

1st–3rd day	1.010–1.012
4th–10th day	1.004–1.008
10th day–6th month	1.004–1.010
6th month–2nd year	1.006–1.012
2nd–8th year	1.008–1.016
8th–14th year	1.002–1.020

High specific gravity as a rule is associated with scanty secretion of urine; low specific gravity with abundant flow of water, and if an unusual quantity of urine is combined with a high specific gravity, the suspicion should at once arise that diabetes mellitus may be present, which is not so rare during infancy as many have supposed. On the other hand, the combination for a considerable time of a large quantity with low specific gravity, must arouse the suspicion of beginning renal disorder, since it is well known that in such a disorder the capacity of the kidney to concentrate the urine is the first thing to suffer.

The *osmotic analysis* has not up to this time acquired either great theoretical or practical importance in pediatrics. The comprehensive examination of Sommerfeld and Röder have shown that the freezing point of the urine is lower in babies than it is in adults, but that it varies between tolerably wide limits according to the method of feeding. The variations are widest in breast-fed babies. The values of the urine of infants must be considered extraordinarily high in comparison with the amount of crystalline matter. Röder and Sommerfeld leave the question open whether the appearance of dissociation is the important factor, or whether the high value is to be attributed to the presence of other, and as yet unrecognized substances. This problem can only be determined by an electrolytic examination. In the infectious diseases, which are often complicated by renal disease, the lowering of the freezing point, and the molecular concentration permit us, even before pathological elements are capable of demonstration, to suspect the beginning of damage to the kidney (Labbé).

The urine voided immediately after birth, is, according to Flensburg, almost clear. Specimens passed later than this are generally turbid, and this condition will continue till the eleventh day. But if the urine does not become clear after the tenth day, we should suspect the presence of some pathological condition. The turbidness is present in urine containing infarcts, as well as in specimens free from uric acid. In the former there are found, according to some authors, not only uric acid and uric acid salts, but also hyaline and epithelial casts. The question as to whether casts appear in the physiological urine of the newborn or not is still undecided.

SECRETION OF NITROGEN

Inquiry into the conditions of nitrogen excretion can have no value unless it is combined with a study of tissue changes. The postulate here stated holds good also in regard to most of the other urinary elements, which are excreted, such as phosphorus, chlorine, calcium, and magnesium, and this fact has only recently found acceptance in pediatric research. Many examiners have concerned themselves with the study of the quantitative excretion of certain elements of the urine at particular ages, without taking any note of the supply of this element, and these examinations are valueless if we accept the postulate advanced in the opening sentence of this paragraph. The ordinary tabulation of the amount of urea-nitrogen excreted by children at various ages may therefore be omitted here. The values given teach nothing unless they are computed with reference to the total metabolism. Under normal physiological conditions, the quantity of nitrogen excreted will depend upon the supply and upon the intensity of cell growth. The more active the cellular development, the more nitrogen will be retained. The character of nitrogen .distribution is of great value in the physiology and pathology of the infant. It is well established that in both infants and older children the greatest part of the nitrogen is eliminated in the form of urea, and that any other conclusion is the result of faulty methods.

The following table from Langstein and Steinitz demonstrates the distribution of the nitrogen in the urine of breast-fed and artificially nourished infants.

Child.	Nourishment.	Daily quantity in cm.	Total nitrogen in 100 ccm.	100 mg. N breaks up into		
				NH_3 — N.	Urea amino-acid — N.	Urea N.
Scholz………… …………………	breast	432.	103.74	23.	72.	69.
Heldt ………………………………	breast	405.	128.3	23.	68.	68.
Bloy ………………………………	fresh milk	222. up to 250.	1292.2	7.	88.	86.
Bloy ………………………………	fresh milk	220. up to 250.	1161.9	6.	88.	86.
Linke ………………… ……	malt soup	165. up to 250.	307.58	3.4	81.	77.
Schāfer…… …………………	malt soup	165. up to 200.	281.7	9.	70.	63.

The quantity of *ammonia*, or the proportion of ammonia-nitrogen to the total nitrogen (ammonia cœfficient) is generally increased in acute disorders of nutrition of infants, while the quantity of urea is correspondingly diminished. This fact has assumed importance in the pathology of infantile metabolism because it shows that the alkalies excreted are not sufficient to neutralize the urinary acids (doctrine of acidosis, see pathology of metabolism). The nitrogen which cannot be precipitated by phospho-tungstic acid, and which cannot be transformed into ammonia by heating with metaphosphoric acid, has been, of late years, called amino-acid-nitrogen. The school of Friedrich Müller has emphatically and justly objected that this nomenclature is unsuitable, if for no other reason, because its results are founded upon an indirect method. Moreover, a considerable proportion of the nitrogen, which is not precipitated by phospho-tungstic acid, and which is not urea, is present in the form of hippuric acid in adults, but we do not as yet know the proportions in which these substances exist in the urine of children. The condition, which has been applied in adult life, that the excretion of amino acids shall be accepted only when the quantities have been directly proved, must hold good when we discuss the matter in connection with childhood.

Recent experiences have proven that under normal conditions, adults excrete amino acids, such as glycocoll, but in infants the presence of this product of albumin-splitting has not yet been demonstrated (investigations by Rietschel at the Berlin Children's Clinic). It is true that there are two well-known disturbances of metabolism which occur in the infant, in which there appear in the urine amino acids or their derivatives: namely cystinuria, and alkaptonuria. The former, which is sometimes hereditary in origin, is generally manifested by the appearance of a sediment consisting of hexagonal tables, which when examined prove to be cystin (aminothiopropionic acid). The crystals are chemically characterized as cystin by the fact that their solution will turn black when boiled with an alkali and lead acetate (formation of sulphide of lead). In alkaptonuria, which is usually congenital (Garrod, Erich Meyer), a derivate of the aromatic group of the albumin molecule, the so-called alkapton, is secreted, which is characterized as homogentisic acid (dioxyphenyl acetic acid). The presence of this acid makes the urine turn black when exposed to the air, a process which is quickened by the addition of an alkali (the mothers state that the urine leaves dark spots upon the baby's napkins), and causes it to reduce alkaline ammonia-silver solution when cold, and Fehling's solution when hot, and to turn green transitorily upon the addition of ferric chloride.

The *excretion of uric acid*, which results essentially from the transformation of the nuclein of the cell-nucleus, is tolerably constant in quantity during the first days of life. Reusing gives the following average figures for the absolute quantitative excretion of uric acid during the first days after birth:—

1st day	0.0410 uric acid in grams
2nd day	0.0411 uric acid in grams
3rd day	0.0831 uric acid in grams
4th day	0.0395 uric acid in grams
5th day	0.0566 uric acid in grams
6th day	0.0463 uric acid in grams
7th day	0.0731 uric acid in grams

It is of interest that in the urine of the newborn there may be found a substance which is closely related chemically to uric acid:—allantoin.

We are indebted to Göppert for a comprehensive study of the uric acid excretion of older children. His results show that it is at present impossible to fix a rule for different ages. Whenever we attempt to draw conclusions from the quantity of excreted uric acid, we must at the same time take cognizance of the diet and of the amount of total nitrogen. It may be stated, by the way, that children from one to two years excrete approximately 0.1 to 0.2 Gm. (1–3 gr.) of uric acid per day (on a mixed diet); from 2–5 years 0.2 to 0.4 Gm. (3–6 gr.); and that the excretion will keep within these limits until the age of puberty.

The following table, taken from Czerny and Keller's Handbook, gives a good summary of the relative proportions of the nitrogen-containing substances in the urine up to the age of puberty:—

	Series.	From 100 total nitrogen are derived					Diet.
		Urea nitrogen.	Ammonia-nitrogen.	Purin nitrogen.	Uric-acid-nitrogen.	P_2O_5.	
Adult, males		85.5	5.0	....	....	20.	mixed.
Youths (14-19 years)	1	80.6	6.2	....	....	19.6	mixed.
	2	83.	5.2	1.69	1.44	14.3	mixed, with plenty of potatoes and bread.
	average	81.8	5.7	....	....	16.9	
Children	3	84.9	7.6	....	....	17.	mixed.
	4	86.9	5.3	1.41	1.18	19.	mixed, with plenty of milk.
	average	85.9	6.4	....	....	18.	
Infants (5½ and 8 months).	5	79.	8.	4.50	....	16.	breast-milk.
	6	84.	5.2	1.05	0.98	28.	cow's milk.
	average	81.5	6.6	2.77	....	22.	

Creatinin has not been demonstrated in the urine of healthy milk-fed babes, but in febrile diseases it may appear, and also when introduced into the system by mouth.

The proportion of carbon to nitrogen is relatively higher in infancy than in adults. While in adults, the carbon-nitrogen quotient will be found to vary between the figures 0.5–0.7, in infancy this ratio may reach 1.0 (Rubner, Heubner, van Ordt). It has been inferred from this that in the urine of infants combinations of carbon and nitrogen are to be found in abnormal quantities, but this supposition is wrong, because the investigations of Steinitz and Langstein have shown that the proportion of carbon to nitrogen is to a remarkable extent subject to alimentary influences, The smaller the absolute quantity of excreted nitrogen, the higher is the quotient of carbon to nitrogen, and vice versa.

The proportions of chlorine, phosphorus, sulphur, calcium, and magnesium excreted have no interest aside from the theory of metabolism. Therefore only a few points will be mentioned here. The amount of chlorine excreted is in the first place influenced by the chlorine supply. The sound organism (or the healthy kidney) answer promptly to an over-supply by a more active excretion. Retention for more than a brief period points to pathological conditions. The importance of retained chlorides as a factor in œdema will be considered in the chapter on nephritis.

Still more complicated are the proportions of phosphorus, calcium, and magnesium. On the whole, the law holds good that the artificially fed baby excretes more phosphoric acid than the nursling. This difference is shown not only by consideration of the acutal figures, but by the proportion of phosphorus to nitrogen. This proportion is in nurslings as 1 is to 7; in artificially nourished infants as 1 is to 2. The quantitative estimation of the phosphates and of the total phosphoric acid is of importance, since their proportion is the measure of the acidity of the urine. According to Keller, the quantity of phosphorus contained in organic combination is absolutely smaller than in adults. A summary of the phosphorus excretion of infants is found in the following table, which recognizes the results of Keller's experiments.

		Quantity of the organic phosphorus	
Kind of diet	Total phosphorus	Absolute	Percentage
Breast-milk	0.0618	0.0061	9.9
Breast-milk	0.0982	0.0081	8.2
⅔ Cow's milk	0.4106	0.0059	1.5

The literature on the subject of the excretion of phosphorus in the urine is still too meagre to permit of any definite conclusions. The so-called *phosphaturia*, which has been more thoroughly elucidated by the work of Soetbeer, Tobler, and others, may be briefly considered. This is a disturbance of the phosphoric acid excretion in elder children, and appears itself in the form of milky, turbid urine, which is cleared up by the addition of acid, and is therefore due to the presence of phosphates. Soetbeer has demonstrated that the primary cause of this disturbance is an excessive excretion of calcium by the kidneys taking on vicariously the function of the intestines, which may be disturbed by some morbid process. The basis of phosphaturia is, therefore, a calcarinuria. Owing to the lack of a sufficient number of reports upon the subject the excretion of the alkali salts has not yet been determined accurately enough in healthy children to be discussed.

The excretion of *sulphur* in infants and in older children has been made the subject of special study by Freund, whose investigations show that the quantity of total sulphur elimination is in direct ratio to the albumin transformation. Freund found, for the sulphuric acid, values of 0.1365 : 0.2171 : 0.1159 Gm. This was in breast-fed babies, while in those fed on the bottle, the quantity was 0.5030 Gm. The absolute quantities of ethereal sulphuric acid were very small, ranging between 0.0091 and 0.0162 Gm. per day. Ponticaccia found values of 1–2 Gm. per day for the total sulphuric acid in healthy older children upon mixed diet, the absolute quantity of ethyl sulphuric acid being in almost every experiment 0.1 Gm.

The elimination of ethyl sulphuric acid is believed to be a measure of the amount of intestinal decomposition, but it must be emphasized that it is of value only so far as the appearance of great quantities of ethyl sulphuric acid indicate an excessive intestinal decomposition, whereas the reverse is not true, since its excretion is affected by many conditions, such as albumin intake, albumin-loss, resorption, etc., which cannot be accurately estimated.

Phenol excretion also is effected by the decomposition in the intestines. F. Meyer has shown that it is less in breast-fed than in bottle-fed babies. He found that in a nursling of six months the daily excretion amounted to 5.87 mg.; in one of ten weeks it was 2.507 mg.; and in the bottle-fed the excretion was considerably higher, about 13.28 mg. per day.

The excretion of *indican*, derived from indol, has been frequently investigated. Friedrich Müller, in spite of contrary statements by other writers, even now defends the opinion that indol does not arise from the breaking up of albumin in the tissues, but results exclusively from albuminoid decomposition in the bowel, of the intensity of which it is a measure. This accounts for the fact that the reaction for indican in the urine is almost invariably negative in healthy breast-fed babies, while it is generally positive in the bottle-fed (Senator, Hochsinger, Zamfiresco). Momidlowski has shown that the excretion of indican is increased in almost every child ill with gastro-intestinal disorder, and that the more severe the intestinal disturbance the greater will be the quantity of indican. In older children, upon a mixed diet, the behavior of indican is the same as in adults. The contention of Hochsinger and Kahane, who maintained that the proportion of indican is abnormally large in tuberculosis, and that it was of diagnostic value in the young, may be considered to be refuted, as may also the observation of A. Mayer that indican is not excreted by atrophic infants, since von Starck affirms that he found increased excretion of indoxyl-sulphuric acid in children suffering from school anæmia.

There have been recorded in the literature some instances of indiguria, with the appearance in freshly voided urine of indigo red.

Urobilin and Urobilinogen.—These are chiefly formed in the intestinal tract, where they result from the decomposition of bilirubin, the billiary coloring matter. But they may also be formed outside the intestines, as for instance in blood extravasations.

According to Giarré, urobilin is completely absent from the urine of nurslings, while it is sometimes found in the bottle-fed. According to statements in the literature, urobilin is often present in the urine of scarlet fever patients, while it is absent in cases of diphtheria. Bookman, who tested these statements in Heubner's Clinic, was led to different conclusions. He studied especially the excretion of urobilinogen in infancy, and arrived at the following conclusions:* The urine of breast-fed infants contains no urobilinogen; that of artificially nourished babies almost always does. Urobilinogen will be found in increased quantity in the urine of infants suffering from intestinal affections, especially in cases where the stools are white (Langstein).

Older children excrete urobilinogen upon a mixed diet in varying amount. Infantile diseases, attended by hæmorrhages, occasionally show increase of urobilinogen. This is true also of paroxysmal hæmoglobinuria. In accordance with the observations of Otto Neubauer, Bookman found that urobilinogen disappears from the urine in cases of obstructive jaundice, if the obstruction of the gall-duct is complete, but that it reappears as soon as the bile once more flows into the intestine. To this degree, Ehrlich's reaction with dimethyl-amido-benzaldehyd is of prognostic and diagnostic value in the jaundice of the young.

The Acetone Bodies.—Acetone, diacetic acid, and oxybutyric acid are formed in cases of inanition, and wherever the carbohydrate metabolism is disturbed. Children are more inclined (Langstein and L. F. Meyer) to acetonuria than adults, and not inconsiderable quantities may be found even in the mildest febrile diseases if they are accompanied by inanition. It is not possible to attribute to this any differential diagnostic importance (L. F. Meyer), and furthermore it gives no idea of the nature of the disease.

Reducing Substances.—It is often claimed that the urine of the young, especially that of infants has a greater power of reducing the metallic oxide than is the case with adults. We cannot, however, draw any far-reaching conclusions from this, because in these investigations the line between healthy and sick children has not been drawn with sufficient clearness. The statements in regard to the nature of the reducing substances are extremely few and very unreliable. Since the proofs offered of reduction are remarkably ambiguous, it should be insisted that the presence of sugar should first have been established by the osazon test, before we can accept the results of a reduction experiment. After warming the urine with muriatic phenyl-hydrazin and sodium acetate for half an hour in boiling water, a precipitation consisting of yellow needles (osazon) will be found immediately, or else after cooling if sugar is present.

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* A reagent for urobilinogen, as has been shown by Neugebauer, is dimethyl-amido-benzaldehyd, which is mentioned by Ehrlich. The reaction may be considered positive when a specimen of urine, mixed with a 2 per cent. hydrochloric acid solution of the reagent, turns red, and presents a characteristic spectrum, when heated after the addition of a few drops of hydrochloric acid. If urobilinogen is plentiful, the red color appears while the urine is still cold.

Binet declares that every normal infant excretes traces of sugar (if the trace is exceedingly small, the osazon test will be negative with diluted urine, and will only be obtained if the urine is concentrated. Therefore if crystals are formed in untreated urine, we may suspect the presence of pathological conditions). Some writers have observed a greater tendency to glycosuria in newborn babes, but it is not certain that these observations were made in healthy infants. Steinitz and Langstein have been able to demonstrate that breast-fed babies with gastro-intestinal disturbance excreted in the urine both milk-sugar and one of its derivatives, galactose. Geelmuyden declares that he found in the urine of diabetic children, in addition to glucose, another kind of sugar (paidose), which was optically inactive. After boiling with acid, these separated out a substance which deflected the polarized beam to the right, and left osazon, with a melting point of 130–175° C. (266–347° F.).

Oxalic acid, as has been shown by the investigations of Parker Sedgwick at Heubner's Clinic, seems to be excreted in absolutely larger quantities by children than by adults. It must be recognized, however, that the mere finding of a sediment of oxalate crystals does not justify the conclusion that there is a greater excretion of oxalic acid.

Diazo Reaction.—Authorities are in accord that in most cases of measles, Ehrlich's diazo reaction is conspicuously positive, although it is true, as stated by Ottfried Müller, that this is not an early symptom, but only makes its appearance after the exanthem has come out. In cases of scarlet fever it was observed frequently by some authors, but rarely by others. Nikos Kephallinós found the reaction positive during the first week in 92 per cent. of the cases of typhoid in children examined by him. In purulent cerebrospinal meningitis he found it negative as often as positive, and he reports that he never obtained it in cases of whooping-cough, erysipelas, mumps, gonorrhœal rheumatism, sepsis, influenza, diphtheria, or syphilis. The diazo reaction is approximately the same in infantile tuberculosis as in the adult form. It is positive in most cases of tuberculosis of the lungs, and of miliary tuberculosis (general). It is only exceptionally found in tuberculosis of the bones and skin, but almost always in tuberculous pleurisy. It is uncertain in meningitis, and in tuberculous peritonitis. "True scrofulosis" is never attended by the diazo reaction, but it is noteworthy that the reaction is strongly positive in a group of tuberculous conditions of the lymph-nodes, which were classed under the heading of pseudoleukæmia. It is therefore possible that the diazo reaction may be of some importance in the diagnosis of tuberculosis, where other clinical evidences of its presence are lacking. Moreover, the reaction was found in no small percentage of cases of lobar pneumonia, but it was nearly always absent in rachitis, in nervous diseases, and in diseases of the digestive, the circulatory, and the genito-urinary systems.

The nature of the diazo body is unknown: it is still even a question whether it is nitrogenous or non-nitrogenous, although, it is true, that the latest, though still unconfirmed investigations, declare that it is a highly complex derivative of albumin. My own opinion inclines to the theory that the diazo body is not a single substance at all, but that in different diseases different substances may produce the reaction. Kephallinós reminds us of the fact that the reaction may even be imitated by the passage of drugs through the urine, and he mentions among the substances capable of producting changes in the color shades, creosote and its derivatives, carbolic acid, thymol, opium, cascara sagrada, and hydrastis. The same may be said of rhubarb and of santonin, which induce the excretion of a pigment body, which turns red in alkaline solutions. It is said that tannin, and tannin-containing substances, when added to diazo-positive urine, will prevent the reaction.

Albumosuria and Peptonuria.—In spite of the elaborate studies on the excretion of albumose and peptone, we cannot obtain a clear idea of the conditions under which they are found in infancy. This much is certain, that in nearly all infectious processes, especially scarlet fever and measles, albumosuria is present, but how far this is the result of the specific infection, how far it depends upon complicating conditions, cannot be discovered from reading the literature, and from this point of view, the whole subject deserves a new study. Siegert states that he has repeatedly observed the excretion of large amounts of albumose in the late stages of scarlatinal nephritis, and he describes the appearance of albumose as a favorable prognostic symptom of speedy recovery from the nephritis.

Toxicity of the Urine.—This is said to be greater in infants than in those of adult age. It appears unnecessary to treat this matter in detail, because the theory of urinary toxicity, which has been advanced especially by French writers, will not bear strictly scientific criticism, because the method is not above suspicion.

ALBUMINURIA

The almost universally accepted dictum that albumin in the urine was always a symptom of disease of the kidney, was not refuted until the celebrated investigations of von Leube in the year 1877. Prior to this the isolated observations, which went to show that occasionally albumin might be found in the urine of otherwise healthy individuals, had utterly failed to shake the general acceptance of the relation between albuminuria and nephritis, which had first been propounded by Bright.

Von Leube has asserted that albumin was present in the urine of 4 per cent. of healthy persons examined by him, and that albumin in 16 per cent. was to be found when the muscles had been previously exerted, and these remarkable results have since been repeatedly confirmed and amplified by a great number of workers.

In examining the urine with the most delicate albumin reagents, the fundamental law has been established that every normal urine contains albumin, and that therefore the excretion of albumin is a physiological phenomenon (Mörner, Posner, Senator). It is true that the albumin normally excreted is in such infinitesimal quantities that it cannot be shown by the common albumin tests (boiling, acetic acid, ferrocyanide of potassium), and this strictly physiological albuminuria has no importance for the practitioner, "for whom a latent albuminuria is of no moment, but only the excretion of albumin which responds to the usual tests, without any preparation of the urine." On the other hand if albumin can be demonstrated in otherwise untreated urine, by boiling or the addition of acetic acid, and ferrocyanide of potassium (it is always best to employ both) the result may be taken to prove the existence of abnormal conditions. The practitioner must now decide the question, which is often difficult, whether the albumin is connected with kidney disease, or whether it is one of the forms of so-called physiological albuminuria, such as may result from the upright position, muscular exertion, fatigue, psychial emotion, cold baths, or diet.

Von Leube has designated all these latter forms of albuminuria as "*physiological,*" "because, we cannot seriously speak of conditions as pathological, when in routine examination the great majority of healthy individuals show more or less albumin in the urine treated by the ordinary tests."

It is not my intention to take part in the discussion which has resulted from this proposition, but for practical motives I have refrained from using in the following discussion the term "physiological albuminuria" in the meaning of von Leube. I understand by the term only that latent excretion of albumin which cannot be shown by ordinary tests.

The physician may meet with two varieties of albuminuria which fall within the range of physiological processes. One is the albuminuria of the newborn which appears almost always immediately after birth, and lasts for a few days; the other is the albuminuria seen in older children due to the upright position, or resulting from a change from the recumbent to the erect attitude. Pavy called the latter cyclic: Stirling, postural: Heubner, orthotic. None of these terms has been generally accepted, and to reconcile the matter, Posner has proposed the name "Essential," while Neukirch suggested "Intermittent Albuminuria."

To avoid repetition it will be convenient first of all to consider the albuminuria of childhood (including those which are the result of diseases of the kidney) from the chemical standpoint.

The albumin appearing in the urine may come from the blood, the kidneys, the urinary tracts, or from other organs. Furthermore it may be albumin taken up directly into the vessels from the alimentary canal, *i.e.*, nutritive albumin. Our present chemical methods do not permit us to separate these different forms of albumin, and to determine their exact source. But concerning the nutritive albumin we are in a slightly better position since it may be identified by the so-called precipitation reaction, *i.e.*, the albumin coming from food, will be precipitated by the blood serum of an animal which has previously been treated by repeated injections of this form of albumin.

Generally we have been content to employ for the urine the division used for blood albumin bodies, and—following the proposal of Hofmeister—to designate as globulin that portion which can be precipitated by partial saturation with ammonium sulphate, and as albumin that portion which is only precipitated by complete saturation. But here we have not taken advantage of the knowledge that blood-globulin is very probably a complicated mixture of several albumin bodies. Instead two divisions have been accepted for clinical purposes, *i.e.*, the so-called fibrin and euglobulin, which precipitate by saturation with 25 per cent. ammonium sulphate, and the pseudoglobulin, which precipitates by saturation with from 25 per cent. to 33 per cent. ammonium sulphate. Although it is somewhat precarious to identify albuminous bodies, one from another by precipitation limits, the matter is so far tolerably clear, but practically it is complicated by the presence in the urine of an albuminous substance which is thrown down by acetic acid, without heating. The qualitative test, which has a certain clinical value, is easily made. Fill two test-tubes to the same height with the urine to be tested, and after adding to both a few drops of dilute acetic acid, shake thoroughly for some minutes. This is necessary because precipitation does not occur immediately. Now dilute with three or four parts of water, and to one of the test-tubes add a few drops of solution of ferrocyanide of potash. When the two tubes are now held against a dark back-ground one can not only determine the presence or absence of albumin precipitated by acetic acid, but can even estimate its proportion as compared to the total albumin. Dilution with water after the acetic acid is added is necessary to prevent the precipitation of uric acid in a highly concentrated urine, which might simulate the albumin sediment. This source of error in the urine of persons suffering with diphtheria has been pointed out by Langer.

Opinion as to the nature of the albumin body precipitated by acetic acid has undergone constant change, and even now its position is uncertain. Mörner, to whom we are indebted for elaborate investigations, regarded it not as albumin, but as a combination (salt) of albumin. F. Müller designated it a globulin, in which both his pupils, Staehelin, and also Oswald agreed with him. Leube and his pupils identified it as euglobulin. Rostoski, who believes it to be exceedingly diffusible, would like to see it excluded from the class of globulins. In opposition stand Obermayer and Keller who consider this albumin body to be nucleo-albumin, *i.e.*, a product of the nuclein substance. This they have inferred from the quality of being precipitated by acetic acid and because it is sometimes possible to demonstrate the presence of phosphorus. The first inference is not conclusive because nucleo-albumin posses the quality of precipitation by acetic acid in common with a long series of other albumins; and the positive proof of the presence of phosphorus can be used in support of the nucleo-albumin nature of an albuminous body, only when phosphorus is detected after its pure exhibition. The detection of phosphorus in an albuminous substance precipitated by acetic acid will always leave room for doubt whether the phosphorus did not come from an admixture, since the extraction of an absolutely pure albuminoid body from the urine is scarcely possible.

My own opinion is that the positive identification of the albumin body is not of much importance, because there remains the possibility that the substance is not always the same chemical entity in the various affections which lead to its excretion. But it was necessary to discuss the tolerably complicated conditions because the albuminuria of the newborn as well as the orthotic form of albuminuria are accompanied by the excretion of the albuminous substance which is precipitated by acetic acid, and because the chemical ambiguity of this precipitate has led to differences in estimating the clinical significance of the symptom.

(*a*) ALBUMINURIA OF THE NEWBORN

The earliest statements to the effect that the urine of the newborn might be albuminous were the result of examinations of the bladder content in still-born children, or in those who perished shortly after delivery. But after Martin and Ruge had shown that urine originally free from albumin became albuminous after remaining in the bladder for 18 hours, previous examinations could no longer be considered conclusive as to the condition of the urine in living subjects. The question whether albuminuria in the newborn is a physiological process was again agitated as a result of the examinations of Dohrn upon living subjects. This author found in 62 per cent. of his cases that the urine discharged immediately after delivery was free from albumin. He found in 32 per cent. distinct traces; in 9 per cent. considerable traces; and in 6 per cent. abundant quantities of albumin. Thereafter the investigations into the presence of albumin in the newborn became more numerous from the combined work of German, French, and English scientists. Flensburg's Table gives a summary of the work of the Germans.

	Flensburg.			Martin & Ruge.			Cruse.			Hofmeier.		
	Heller's reaction.			Reaction not mentioned.			Boiling, Heller's reaction.			Reaction not mentioned.		
Day.	Number of examinations.	Number of cases of albuminuria.	Albuminuria per cent.	Number of examinations.	Number of cases of albuminuria.	Albuminuria per cent.	Number of examinations.	Number of cases of albuminuria.	Albuminuria per cent.	Number of examinations.	Number of cases of albuminuria.	Albuminuria per cent.
1st	56.	27.	48.	17.	5.	29.	..	..	..	15.	14.	93.
2nd	25.	10.	40.	17.	7.	41.	9.	5.	55.	9.	8.	89.
3rd.,....	24.	3.	12.	17.	6.	35.	10.	6.	60.	12.	9.	75.
4th.....	22.	3.	13.	17.	3.	17.	10.	1.	40.	11.	9.	82.
5th.....	23.	4.	17.	17.	4.	23.	10.	5.	50.	6.	3.	50.
6th.....	14.	1.	7.	17.	2.	11.	10.	4.	40.	8.	4.	50.
7th.....	20.	1.	5.	17.	1.	6.	10.	2.	20.	6.	2.	33.
8th.....							10.	1	10.	..	..	..
9th.....							10.	1.	10.	..	..	..
10th.....							10.	0.	0.	..	..	..
12th-14th							..	..	..	..	..	..
Sum.....	184.	49.	26.	119.	28.	23.	89.	28.	31.	67.	49.	73.

From this table we learn that in a large percentage of newborn children albumin appears in the urine after delivery, and that it persists for a longer or shorter time but under normal conditions it generally disappears by the tenth day. The French scientists arrived at different conclusions. Parrot and Robin declare that they never have been able to prove, even with the most delicate methods of examination, the presence of even traces of albumin in the healthy newborn ("chez les Enfants bien portants"). Throughout the French literature the opinion prevails that the investigations which showed the occurrence of albuminuria in the first years of life, do not add anything to the physiology of the urinary excretion in the newborn.

It may now be interesting to consider the theories which have been advanced to explain the occurrence of this albuminuria. We find mentioned as *causes:*—"Functional disturbance appearing after delivery, with resulting renal hyperæmia; defective development of the glomeruli in the newborn; albuminuria in the mother; and infarctions of uric acid." There seems no doubt that, among these causes, uric acid infarction has some significance, because its coincidence has been shown by numerous examinations. But on the other hand, it should not be forgotten that albumin has often been found where there was no infarction: often enough indeed to disprove the general validity of this parallelism, in which Hofmeier was inclined to believe.

Infarction, according to Flensburg, induces the appearance of albumin by mechanical irritation. But we may readily conceive a chemical irritation, such as appears in adults as the result of high concentration and an increased excretion of uric acid. Casts may also be explained by infarction producing mechanical and chemical irritation.

We are indebted to Ribbert for the study of the anatomical conditions which are the basis of the albuminuria of the newborn. According to this author the epithelium of the urinary tracts does not play an essential rôle. He believes that the whole of the albumin is derived from the glomeruli, because he was able to prove coagulation in their capsules in the newborn by fixing with alcohol and boiling water. However, according to Ribbert, not only is the albuminuria of the newborn a continuation of the embryonic process (it is well-known that the glomeruli of the fœtus excrete a permanently albuminous fluid), but the increased metabolism of the infancy he regards as one of the most important causes of albuminuria.

In our efforts to explain the albuminuria of the newborn we are still far from escaping mere hypotheses, and as long as this is true all discussions to determine whether or not the albuminuria is a physiological process or not are of little value. Czerny and Keller believe that it is absolutely necessary to a profitable discussion of this point that the relations between nutrition and processes in the gastro-intestinal tract on the one hand, and the albuminuria on the other should have first been studied.

To-day this much is certain—and it has a practical value—albuminuria in the first days of life is of no serious significance, nor is its anatomical substratum by any means an inflammatory process in the kidney. It is only when albuminuria persists and can be detected by the ordinary tests after the tenth day that we have to do with a condition which really is normal.

It seems to be necessary to enter briefly into the discussion of the quality and quantity of the excreted albumin. Flensburg identified the albumin with nucleo-albumin, and added these words:—"This albumin body has not yet been proven to exist in the blood, but only in the urinary tracts and in the kidney substance." What has already been said relieves me of the necessity of emphasizing the fact that Flensburg's remarks not only as to the genesis, but as to the nature of this albumin body, are purely hypothetical. It no more admits of proof than the statement made many years ago, and recently revived, that this albumin is not blood-albumin at all, but mucin from the urinary tracts, and that therefore we have no right whatever to speak of an albuminuria, but only of a mucinuria. In refutation of this, I am able to state as the result of a great many examinations of my own, that the albumin body precipitated by acetic acid is almost always to be found in the urine of the newborn, but that in addition there is present in most cases an albumin which we may designate as either albumin or globulin according to the limits of precipitation. According to my experience the quantity of albumin excreted was from 0.2 per cent. to 2.0 per cent.; the proportion of albuminoid precipitated by acetic acid to the total albumin varying.

(*b*) ORTHOTIC ALBUMINURIA (Heubner).

(Synonym: Cyclic Albuminuria, Pavy)

Orthotic albuminuria is that form of albumin excretion which occurs in an affected person on assuming the upright position and is dependent on the change in posture.

From the time that Pavy, Bull and von Noorden published their first observations on this affection, the literature has attained considerable dimensions, Germany being comparatively late in following the advances of the English and French profession.

The affection preponderates in the period from infancy to puberty. Children whose night urine is free from albumin, excrete albumin in the course of the day. It disappears completely in prolonged decubitus and reappears immediately as soon as the upright position is assumed. Thus there are no external causes responsible for this condition, but the external factor of changing the horizontal for the vertical position. It is associated with the ordinary conditions of life and manifests itself in the fact that the albuminous content of the urine occurs and increases during the day and that it decreases or disappears at night. Continued erect position therefore gradually loses its specific significance, since the cycle can be influenced at will. Heubner's term "orthotic" is therefore preferable to Pavy's designation "cyclic." It was Sterling who, in 1887, first recognized that the erect position occasioned albuminuria and for this reason he termed it "postural albuminuria." Teissier selected the term "Albuminurie de la station debout," or "albuminurie orthostatique." The reason why Heubner employed "orthotic" instead of "orthostatic" is that the latter term merely expresses the act of assuming the erect position.

Albuminous Excretion in a Qualitative and Quantitative Respect.—Nearly all authors who have investigated the chemical nature of the albumin excreted in orthotic albuminuria, notably von Leube, Dresser, Keller, Oswald, Rostoski and Cloetta, have been struck by the fact that the albuminous substance which was present either alone or together with others, could be precipitated by acetic acid. This fact has revealed interesting relations between orthotic albuminuria and that form of albumin excretion which von Leube observed in his wholesale examinations of soldiers after drill. He found that if the drill was light, there was nucleo-albumin in previously non-albuminous urine, while after strenuous service there was also sero-albumin.

In a large number of quantitative examinations in orthotic albuminuria I have found three types of cases: (1) in which none but the albuminous body is excreted which is precipitable by acetic acid and which is generally associated with slight albuminuria; (2) in which not only the albuminous body, precipitable by acetic acid, is excreted, but also true proteid, the quantity of which may amount to more or to less than that of the former; (3) in which all three kinds of albuminous substances —the one precipitated by acetic acid, albumin and globulin—are excreted in varying proportions. This division requires only some limitation in the sense that cases in which one or two kinds are evacuated have been but rarely observed, and if I refer to their occurrence at all, it is merely to say that more than one proteid substance were present in quantitatively undeterminable traces.

The proportion of the substance precipitable by acetic acid to the total quantity of excreted albumin was about 30 to 100 in twelve cases, 90 to 100 in the great majority, and from 7 to 16 in a minority of seven cases. In 78 per cent. of all cases the quantity precipitated by acetic acid exceeded that of pseudoglobulin, and in 22 per cent. it amounted to less.

These results are not only of value for healthy persons after exercise, but also, and even in a higher degree, when compared with those forms of albuminuria which are symptoms of acute or chronic nephritis. In this connection it is of importance to note that among the chronic renal diseases, which alone can give rise to a differential diagnosis with orthotic albuminuria, amyloid kidney is the only one which is accompanied by the excretion of large quantities of albuminous bodies precipitable by acetic acid, while only small quantities, if any at all, are present in so-called chronic nephritis of both adults and children.

What does the urinary excretion of albuminous bodies reveal as to the nature of the process on which orthotic albuminuria depends? Regarding it as nucleo-albumin, as did von Leube, Obermeier and Keller, and regarding nucleo-albuminuria as the expression of degenerative changes of the renal epithelia ("Mauserung"), we have to infer from its appearance the presence of renal changes in orthotic albuminuria. This theory, however, which is held by Keller, is not free from objection even without calling to aid an exact investigation of the nucleo-albuminous nature of the proteid body. In the first place, the latter is absent in chronic affections of the kidney, although the large quantities of desquamated epithelia in the urine furnish unquestionable proof of degenerative processes. In the second place, it is often excreted through the urine for months or years, so that the renal epithelia would not be able to supply the necessary material. We unreservedly share von Leube's opinion that it passes direct from the blood into the urine. Even though the correctness of Rostoski's opinion that this substance is of an easily diffusible nature, may be questioned, it nevertheless seems to be a fact that its excretion bears a different significance to that of other albuminous bodies, and the separation of that form of albuminuria to which alone it is confined seems to be the most plausible proceeding (Raudnitz, F. Kraus).

The absolute quantity of the excreted albumin may, according to my experience, vary very widely. It may be "minimal," but it may also attain to the high values which occur in the most severe renal affections, amounting to as much as from 2 to 5 per mille. It varies in the same individual under apparently unchanged conditions of life, and its intensity depends upon factors as yet undiscovered.

Occurrence.—In 1890 Heubner stated in his well-known work that orthotic albuminuria was of rather rare occurrence, but this opinion, to which he still adhered until 1897, may now be considered obsolete. All those who have a large juvenile material at their disposal upon which to make systematic observations agree that albuminuria is of frequent occurrence. This conviction originated in France, although a few French authors discredited it, Teissier for instance as late as 1905. In a large number of cases Leroix found, in 1883, one case in every seventeen children. Langstein and Reyher, of the Royal Charity Hospital, found orthotic albuminuria in 12 per cent. of all admitted children. Martin found albuminuria in 86 out of 304 debilitated children, or 38 per cent., Jehle 39 per cent. in 223 carefully observed cases.

According to Heubner's statistics the frequency of the affection is distributed over the various periods of life as follows:

From 0 to 15 years.	22 cases, or 39 per cent.
From 16 to 20 years	21 cases, or 38 per cent.
From 21 to 30 years.	10 cases, or 17.86 per cent.
Over 30 years.	3 cases.

Schaps, who made investigations at the Breslau Clinic, found 91.12 per cent. between the ages of 5 and 15. All authors are agreed that the affection is exceedingly rare in early childhood and that its greatest frequency sets in toward puberty. Schaps found only 1 case (2 per cent.) under 5 years; 15 cases (41 per cent.) from 5 to 10 years; 18 cases (53 per cent.) from 10 to 15 years; and only 1 case (2.9 per cent.) over 15 years.

Langstein saw 2 cases of 3 years, 3 of 4 years, 2 of 5 years and 5 of 6 years. It was only after 6 years that he observed increased frequency. In Jehle's statistics the frequency of the affection is tabulated as follows:

From 0 to 6 years among 35 cases	1, or 2.8 per cent.
From 7 to 10 years among 111 cases	32, or 19.8 per cent.
From 11 to 14 years among 84 cases	59, or 61.2 per cent.

Opinions are divided as to the susceptibility of the sexes. Dubreuille places the number of male patients at eight times larger than that of females; Oswald places the proportion of males to females at 4 to 3; Heubner found that the female sex suffers to a vastly greater extent from orthotic albuminuria than the male. Klemperer and Schaps are of the same opinion and personally I must also share it, since there are 66 female cases in 87 own observations. On the other hand, Jehle did not observe any particular difference, finding 45 among 107 girls and 42 among 116 boys.

As a family affection orthotic albuminuria was first described by Heubner, later by Rudolf, Hoxon, Schoen, Lacour, Schaps, Langstein and Jehle.

Symptomatology.—There is no need to further discuss the symptoms that dominate the clinical picture and gave the affection its name. The picture, however, has not yet been characterized with the desirable degree of accuracy, which is partly explained by the fact that the various authors had not immediately seized upon the correct conception of the disease. Thus, a number of investigators spoke of orthotic or cyclic albuminuria as an affection *sui generis* where, aside from albumin, hyaline casts, epithelial casts and renal epithelia were demonstrable in the urine (Keller and Stridsberg). Although the value attributed at the time to the presence of casts in the urine for diagnosing renal affections has depreciated during the last few years, an important aid to differential diagnosis would be sacrificed, if the results of the examination of the sediment were to be ignored. It has certainly been established that the presence of a few isolated hyaline casts does not justify the diagnosis of a renal affection, but it would be well if even at the present time the presence of epithelial and granular casts, aside from that of renal epithelia, were regarded as the expression of renal involvement, and the conception of orthotic albuminuria confined, in conformity with Heubner, to such narrow limits as to refer to cases which show the typical behavior of albumin secretion to the exclusion of those whose urine contains casts and other renal elements. Jehle's recent investigations, to which I shall refer later in detail, seem to show that the presence of even large numbers of casts and other form elements does not justify the exclusion of a diagnosis of orthotic albuminuria. On the other hand, Goetzky's extremely careful, numerous examinations of the sediment of the urine of children with orthotic albuminuria have shown that this affection runs its course without the excretion of form elements, and casts in particular. Goetzky obtained on many successive days each portion of urine, both albuminous and non-albuminous, and examined the freshly prepared sediment. Where casts were present, even in small numbers, he also found other deviations in the urinary picture and anomalies in the albuminous excretion, which would render it doubtful whether there was true orthotic albuminuria. It would be wise, therefore, to take Heubner's words to heart:

"The presence of well developed hyaline casts, especially the cellular and granulated varieties, or of epithelia, should as a measure of precaution always be looked upon, at least in children, as a sign of nephritis and not of orthotic albuminuria."

A few cases are mentioned in the literature where from time to time the morning urine contained albumin. Although it is possible that this constitutes a different form of juvienile albuminuria, it should be considered that the cycle may have been disturbed from insufficient evacuation of the evening urine and from children having urinated during the night. Otherwise the urine of true orthotic albuminuria does not present any noteworthy peculiarities. Its often turbid consistency is attributable to the presence of phosphates or oxalates, the latter being particularly frequent in some cases. The sediment contains but very few round cells, also some mucus or mucous cylindroids, the significance of which is open to doubt. Sometimes there are seminal threads in the urine of boys at the age of puberty or, as Heubner emphasizes, fragments of a substance which probably emanates from the prostate which might be mistaken for cast fragments. In girls nearing puberty the urinary sediment contains with great regularity numerous vaginal epithelia which point to desquamative vulvitis. They will, according to my experience, disappear with the first menstruation.

Abnormal excretion has been accidentally discovered in children who felt absolutely healthy. In the great majority of cases, however, there are manifestations which point to a disturbance of the general condition. Heubner characterizes these symptoms as follows: "There is considerable atony and lassitude which deprive the children of all joyfulness, alacrity and pleasure of work, or there is headache, articular pains and symptoms of weakness."

Gillet distinguishes between three forms of the clinical picture. Some of the patients have no complaints whatever, others have widely varying functional disturbances (dyspepsia, neurasthenia, adolescent pains), while the third group has headache, transient œdema and pains in the sacral region. It is an easy matter for me, too, to pick out three classes out of the number of cases I have observed: (1) Those with a pale complexion, tendency to fatigue, palpitation, anorexia, headache: complaints which are associated with chlorosis; (2) A smaller number of children looking flourishing, but complaining of headache, congestions to the head, occasional vomiting, colic, recurrent urticaria; (3) The third group having no pathological symptoms at all, where the presence of albuminuria was discovered by accident.

The objective findings of the albumin have long been neglected, with the exception of its periodical coming and going, more stress having only recently been placed upon them, notably upon the examination of the heart and vascular system. But these are the only points in which abnormal conditions have been observed. Schaps, for instance, reports having observed more or less pronounced pathological cardiac manifestations, such as palpitation, dicrotia, strong apex beat, arrhythmia and noises, in 20 patients out of 35. He has also observed dilatation of the heart which affected either the right or the left side and repeatedly alternated. Generally speaking, the symptom-complex observed by Schaps in regard to the circulatory system, agrees with that described by Germain See under the name of "hypertrophie et dilatation de la croissance." Schaps having observed but one case with cardiac dilatation, hesitates to describe its nature. Stridsberg positively demonstrated hypertrophy of the heart in 13 among 31 cases, the symptoms in the remaining cases being uncertain. Lommel, Krehl and Loeb have observed about the same cardiac manifestations as Schaps. Krehl summarizes the cardiac changes under the term "cor juvenum." I have not been able, with the material at my disposal, to find such a large number of cardiac anomalies as Schaps, although I have occasionally found increased frequency of pulse, arrhythmia and systolic noises at the apex. Considering that some authors, Stridsberg among others, have unquestionably included cases of renal affections, it may well be supposed that the results of cardiac observations are not yet ripe for comparison. Reyher made orthodiagraphic examinations of the hearts of children with orthotic albuminuria and found that in the great majority of cases there was arrest of heart development ("Tropfherz"). In one case an electro-radiogram was taken by Heubner which showed the presence of the infantile prong which is normally present in the first half of the period of childhood, while in adults it occurs in nervous, neurasthenic patients. "These findings point to the continuance of a certain infantilism in the region of the circulatory organs, a nervous disturbance of the cardiac function." Otherwise, signs of cardiac weakness could not be discovered in the diagram.

Matthes, like myself, has never observed any particular tension of the pulse or elevated blood pressure. In a few cases the blood pressure was determined by Heubner with von Recklinghausen's apparatus, with the result that in patients in a reclining position the systolic pressure was 140, 150 up to 160—figures which, although not pathologically high, were by no means unusually low. The pulse pressure was usually low, because the diastolic pressure was high. The fundus of the eye was always found normal with the exception of the case I am about to report in detail.

In the majority of cases the hæmoglobin content was found normal, even in children with unusually pale complexions. The latter was therefore not the result of anæmia, but of an abnormal distribution of the blood, or angiospasm.

Pathogenesis, Nature, and Etiology.—There is no dearth of theories to explain the presence of orthotic albuminuria, but in their totality they are more or less hypothetical. They may be conveniently divided into two groups: one regarding an affection of the kidney, with anatomical structural changes, as the etiological factor, the most prominent supporters of which are Johnston and Senator; and the other thinking that the affection has nothing to do with an impairment of the renal tissue, as supported by Heubner, Posner, Ibraham, Neukirch and others. One of the reasons why this difference of opinion has lasted to the present day is that there was no autopsy case on which to study the pathological anatomy. I am fortunate enough to fill this gap by the study of a case which was observed by Heubner and myself from the beginning of the affection until its fatal termination and in which the kidneys were histologically examined in the most painstaking manner. The course of the case appears so important for the ventilation of the whole question that its detailed communication at this place may be worth while.

Agnes K., ten years old, was admitted to the Berlin University Children's Clinic on March 10, 1903. A noteworthy point in the history is that the mother has aborted ten times. The child knew nothing of a syphilitic affection, and any guiding point in this respect could at first not be obtained. One sister died of tuberculosis, one of scarlet fever, while one brother is healthy. Had measles at the age of six months, the case being attended by a physician; at the age of seven there was bronchitis; at the age of eight chickenpox. In July, 1903, she contracted a cough which improved after a stay at Pyrmont, but afterwards recurred. The Pyrmont physician could find nothing wrong with the child who came to the clinic owing to her cough and a slight glandular swelling of the neck.

Examination revealed nothing particularly abnormal but for a few palpable cervical glands. They were from pea to bean size. She had a flourishing complexion, red conjunctivæ and mucous membranes, was intelligent and active. There were a few crepitant râles over the right lung, the heart was not pathologically changed and the urine was free from albumin. Hydropathic measures improved the bronchitis after a slight febrile exacerbation; the glandular swellings receded upon mercury inunction and iodine treatment. During the entire time of observation there was no albumin present in the urine, until, on May 24, 1904, the child again visited the clinic owing to headache and vomiting. The urine was then found to contain the unusually large quantity of almost 4 per mille of albumin, in which the substance precipitable by acetic acid predominated. Careful clinical observation during the next few days, together with the quantitative analysis of the various portions of urine, showed that this was a typical case of orthotic albuminuria, in which the excreted albuminous quantities varied from day to day. Psychic excitement was found to lead to considerable increase of the albumin. In spite of repeated thorough examinations no casts could be found.

Owing to its considerable interest, the case was admitted into the clinic, where clinical observation established the fact that the case was one of orthotic albuminuria. Metabolic experiments, made for over a week, elicited the fact that the quantity of uric acid was abnormally large in proportion to the excreted nitrogen and the daily excreted quantity of oxalic acid was between 50 and 70 mg. Crystals of the latter were found in the sediment. When the child was discharged, the suspicion of nephritis was excluded.

I had not seen the child until the beginning of November, 1904, when she was sent to me by an ophthalmologist with the diagnosis of retinitis albuminurica, associated with nephritis, and I frankly confess that I was not particularly pleased with the position of affairs. The child having complained about glittering before the eyes, the mother had obtained an oculist's advice. There was headache and vomiting. Upon thorough examination I found no symptom to induce me to agree with the diagnosis of the oculist. Heart and vessels were normal. The daily urine certainly contained large quantities of albumin, but no casts. Ophthalmoscopical examination revealed pronounced choked disc and perivascular choroiditic foci at the periphery of the fundus. No doubt this picture could simulate retinitis albuminurica, but as the child's appetite was excellent in spite of vomiting, and dyspeptic disturbances which belong to the picture of uræmia were completely absent, I refused to accept the diagnosis of nephritis with uræmic symptoms. Von Michel gave an opinion of bilateral choked disc with choroiditic foci at the periphery of a luetic nature. A provisional diagnosis was made of cerebral tumor, possibly of a syphilitic nature, and orthotic albuminuria. The further course confirmed the diagnosis of tumor, but antisyphilitic treatment was unsuccessful. Ataxia of the extremities and blindness followed next. Then came paralysis of the ocular muscles, spasms of the extremities, a pronounced alteration in the shape of the head, hydrocephalus rapidly developing which rendered an exact topographical diagnosis of the tumor impossible. While the patient remained in the dorsal decubitus, the urine was always free from albumin which, however, appeared even during the last few months whenever she sat erect outside the bed. Casts were never found. Gradually there was periodical unconsciousness alternating with epileptic paroxysms, ending in death on October 10, 1905, during an epileptic attack.

Autopsy revealed a cerebellar tumor, gliosarcoma of the left hemisphere, and a considerable hydrocephalus. There were tuberculous foci in both apices of the lung, the heart was quite normal, and the abdominal organs were extremely plethoric. The kidneys were not enlarged, the capsule was easily detached, the surface thoroughly smooth but for a contraction the size of a pea at the surface of one of the kidneys. Macroscopically, the renal section was absolutely normal. The most painstaking microscopical examination (Heubner) established absence of any inflammatory or degenerative changes. The loops of the glomeruli were perfectly tender, transparent, in several places well filled with blood corpuscles and free from any exudates whatever. "Sections 5 mm. thick showed that there could be no question of a nuclear increase either of the capsule, glomerular epithelium or capillaries. Neither was there any exaggerated number of leucocytes in the capillaries." The contracture was occasioned by a relatively fresh infarct, probably owing to a marantic thrombosis of a small vessel which had recently developed. A few epithelia of the uriniferous ducts in this region showed slight fatty degeneration.

The importance of this case requires no comment. Had the mother contented herself with the oculist's diagnosis it would have been impossible to observe all the details of the case to the end, and the case would have served as a support for the opinion of those who would derive orthotic albuminuria from an affection of the renal tissue. This autopsy has brought out the fact that there is no such connection and that true orthotic albuminuria may occur independently of any anatomical change of the kidneys.

Among the theories which primarily inclined to this view, the cardiovascular one will probably have the most supporters. Schaps' opinion that the anomalies of the circulatory organs leading to orthotic albuminuria belong to the group of "hypertrophie et dilatation de la croissance" has recently found a new supporter in Loeb, who studied the behavior of Korany's quotient $\frac{\Delta}{\mathrm{NaCl}}$ in individuals with orthotic albuminuria and concluded from his investigations that orthotic albuminuria is of a circulatory nature. According to Korany the increase of the quotient admits of the conclusion that there is diminished speed of the blood current of the glomeruli and consequently the kidneys, while a decrease of the quotient proved increased blood circulation in the vessels. It may be taken that this idea has been pretty generally accepted as correct. Loeb found in his patients that with the onset of albuminuria there was always a decrease of urine and nearly always increased concentration. In all cases, however, there was an increase in the quotient. Nephritic patients did not react in the same way. Those with good cardiac function went through the experiment of rising without any considerable increase in the quotient, while those with cardiac insufficiency were affected like patients with othotic albuminuria. Moreover, experiments made by Knecht with cardiac insufficiency showed that "not the affections themselves, but cardiac insufficiency causes the result in the experiment of rising from the decubitus." In this way Loeb comes to agree with Edel in making orthotic albuminuria exclusively dependent upon circulatory disturbances of the blood current. On the other hand Leube had previously explained that it is impossible to ignore the assumption of individual degrees of congenital permeability of the renal filtration membrane. He believes that the renal filter is not equally permeable in all individuals, and in illustration for his hypothesis cites the family occurrence of orthotic albuminuria. According to von Leube the human race is divided into 3 groups: (1) those with an absolutely permeable renal filter who excrete albumin under normal conditions; (2) those with a relatively permeable filter who excrete albumin under the influence of physiological conditions of life; (3) those with a relatively impermeable filter who will not react with albuminuria to the conditions referred to.

Another theory, which is supported by Sutherland, Mosny and Goublain, is that orthotic albuminuria is not dependent upon circulatory disturbances of the blood current, but upon those of the renal vessels, laying stress upon the connection between this affection and floating kidney. But as this is a very remote coincidence, the theory can for this reason alone have no general validity.

The oft-repeated assertion that there is a connection between orthotic albuminuria and previous infectious disease does not seem quite clear. The idea is that these diseases should have resulted in a "weakening" of the renal vessels, thereby leading to the occurrence of the anomaly. This, however, is refuted by the fact that this connection is absent in a number of cases and that there is no proof whether the cases cited in support of this theory did not have typical orthotic albuminuria previous to the contraction of the infectious disease, and furthermore, that probably the majority of cases in which nephritis occurred after an infectious disease, were simply in the last phases of nephritis which must be rigorously separated from those of essential orthotic albuminuria.

Neukirch supports the nerve theory, holding disturbances of the renal innervation responsible for orthotic albuminuria. Teissier blames the destruction of red blood corpuscles following hepatic functional disturbances as the causative factor. Ralfe, too, inclines to this opinion, having observed orthotic albuminuria in individuals who had previously suffered from paroxysmal hemoglobinuria. According to another hypothesis there is a connection between albuminuria and exaggerated demand upon the cells of the marrow, while von Noorden believes that there is a general metabolic disturbance which is as yet unexplained. I have myself inclined to the latter opinion, having found unusually large quantities of oxalic acid in the urine of some cases, but inasmuch as the latter is not restricted to orthotic albuminuria, the assumption of a metabolic irregularity can no longer be held to support this contention.

Méry and other French authors agree with Froehlich in distinguishing between a renal form of intermittent and functional albuminuria. The renal form is said to be a deuteropathic manifestation of acute nephritis and to be carefully distinguished from chronic nephritis. Méry's definition of this form is: "The symptoms of chronic nephritis are absent, but bloatedness, headache, and tense pulse are present." According to this author the functional form is intermittent from the beginning; the complaints of patients are of a general nature; at the same time there are frequently vasomotor anomalies; exaggerated exercise and nervous excitement increase the albuminous excretion. Méry emphasizes the urological cycle of Teissier as a characteristic peculiarity of the functional form: "The manifestations consist in the fact that in the daily urine there is successive or increased secretion of pigments, albumin and uric acid, which daily recurs in definite cycles." From an etiological point of view Méry divides the functional form as follows: the gout-threatening form, which is the most frequent and important and mostly affects neurasthenic, over-fed individuals; the hepatogenous form, which is often associated with acholuric icterus; the digestive form, which is characterized by the excretion of (nutritive) proteids during the digestive period exclusively; the mechanical form, which may be of an orthotic or a hypostatic type; and the pretuberculous form.

It will be seen from the foregoing that the varying conception of orthotic albuminuria is not only exemplified by the description of the symptomatology, but also in attempts at its explanation. Thus, the renal form of the French evidently comprises only forms of terminating nephritis, and it may have been just this kind of cases in which pronounced involvement of the heart or vascular system has been observed.

The pathogenesis of orthotic albuminuria can only be thoroughly understood if two preliminary questions can be satisfactorily answered. They are:

1. How does it happen that anatomically intact kidneys secrete albumin?

2. How can it happen that the factor causing the albuminous secretion is only active in the erect position of the body?

Let us first examine in how far these two questions have already been answered by the theories described and by those which will still have to be described.

Albuminous excretion as such may be occasioned by albuminous substances coursing in the blood which cannot be assimilated by the organic cells. Von Noorden, among others, at the time seriously discussed the possibility whether the pathogenesis of orthotic albuminuria might be thus explained. The theory could be supported by the behavior of the excreted albumin, since it showed the property of being precipitable, wholly or in part, by acetic acid. At any rate, there is no orthotic albuminuria without the excretion of albuminous bodies that can be precipitated by acetic acid. Since, however, this property characterizes but very imperfectly the nature of the protein substances it is clear that no conclusion can as yet be drawn from it in regard to the quality of the excreted albumin, and no discussion can be profitable as to whether it is assimilable or not. Besides, the fact of insufficient assimilability would only explain the albuminuria, but not the phenomenon of orthosis.

It will be necessary, therefore, to look for other explanations.

Von Leube's idea of dividing the human race into individuals with permeable, relatively and absolutely impermeable renal filters, is of little value, since the assumption that the urinary substances are secreted by a filtration process is no longer tenable. True, Runenberg has shown that under low pressure proteid solutions can be better filtered than under high and is inclined to refer albuminuria to reduced blood pressure. Heidenhain has pointed out that experiments made with dead membranes are no criterion for processes going on in the living renal parenchyma; besides, he gave Runenberg's experiments a different interpretation.

It would materially advance our knowledge of this theoretically and practically interesting form of albuminuria, if it could be decided whether the renal epithelia become permeable to the protein bodies of the blood, or whether protein substances are secreted from the cellular structures into the urine. It is assumed that toxic injury to the renal epithelia or kinking of the renal artery leads to excretion of nucleo-albumin from the nuclear substances of the renal epithelia. True, the albumin excreted in orthotic albuminuria shares with nucleo-albumin the property of being precipitable by acetic acid, but, so far as present investigations admit of a conclusion, it contains no phosphorus. It may, therefore, be permissible to assume that the "functional" lesion of the renal epithelia which must always be present in albuminuria, has rendered the epithelia permeable to the proteid of the blood; and since the epithelia of the kidney pre-eminently show this peculiarity in circulatory disturbances, the idea suggests itself that the latter, or vasomotor insufficiency, are responsible for orthotic albuminuria.

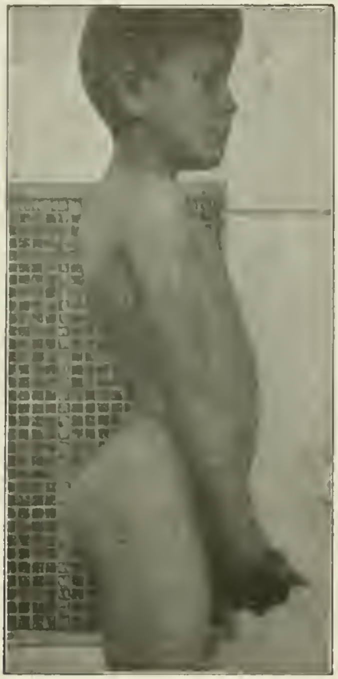
Edel was the first to make this suggestion, but he failed to explain what peculiarities of the cardiac and vascular functions will cause albuminuria when changing the horizontal to the erect position. He pointed out, however, that factors favoring the disappearance of albuminuria also lead to changes in the pulse, which in volume and frequency was more resistant to certain demands in non-albuminous than in albuminous periods. I have pointed out the great frequency of vasomotor phenomena in a great number of orthotic children and differentiated between an angiospastic and erethic type, according to the preponderance of the vascular spasm in the clinical picture. Martius observed cardiac dilatation and weakness in a very large number of patients, although other investigators noticed considerably fewer such cases or none at all. Increasing experience with orthotic albuminuria tends to support the opinion that this affection is connected with the function of the circulation. Experiments made by Erlanger and Hooker, as well as by Frank, explain this connection.

Erlanger and Hooker made their experiments on two young men, aged 29 and 28 respectively, the former being healthy and the latter having been subject to orthotic albuminuria for five years when the condition was first observed. Blood pressure, pulse pressure and circulation were determined by precise instruments under varying conditions. They let the orthotic patient breathe against high air pressure in the decubitus, in order to increase the venous pressure in the general circulation and found the urine free from albumin. The same findings resulted when in the horizontal position the intra-abdominal pressure was raised by tension of the muscular abdominal wall. Erect position at once produced albuminuria, but failed to appear if the body weight was eliminated by immersion of the erect body in water. American investigators have demonstrated the fact that elimination of muscular exertion and tension does not prevent albuminuria. In a sitting posture violent movements with arms and legs did not lead to albuminuria, while the latter, when present, was lessened by walking about.

The orthotic patient commenced to excrete albumin when erected to an angle of about 40°, but this did not occur when the patient was turned in the reverse direction. This experiment rendered it probable that albuminuria was caused by insufficient regulation of the level of the total organic fluid, the lower extremities being insufficiently supplied; and this pathogenesis became almost a certainty when no albumin was secreted upon compressing the lower extremities by pneumatic stockings which drove the blood and tissue fluids upward under a pressure of 50 mm. Hg. The pressure measurements, taken in a large number of experiments, showed that concurrently with the onset of albuminuria there were: increase of the minimal pulse pressure, decrease of the pulse pressure, increase in the pulse frequency, unchanged product of pulse frequency and pulse pressure combined.

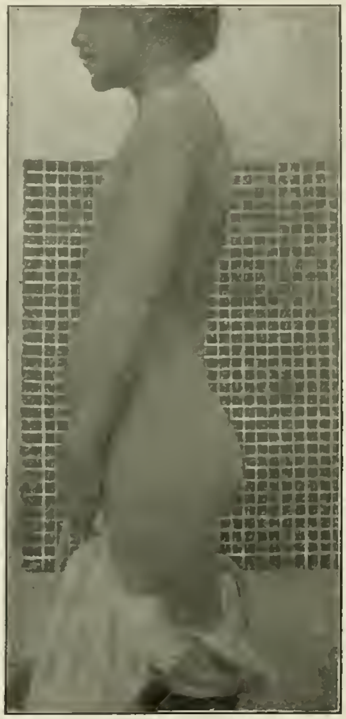
Frank, too, thinks that the chief factor in orthotic albuminuria is an injury to the renal elements owing to deficient blood supply. He considers his view supported by the established fact that albuminuria is always associated with oliguria. Diuresis is even impaired, at least to a certain degree, under the influence of diuretics. Albuminuria disappears in the erect individual under the influence of faradization at any part of the body. Frank believes, as a result of his highly interesting experiments, that the vascular disturbance of the kidney is a reflex process.

Fig. 2a.



Characteristic lordosis in a boy of 12.

Fig 2b



Normal curvature of the vertebral column in a girl 12 years old.

Erlanger and Hooker had previously pointed out that the position of the erect body had a certain influence upon albuminuria, the latter decreasing when patient bent forward over a table at right angle. Frank amplified this statement by demonstrating that, aside from lying down or sitting, standing up with the body bent forward caused albuminuria to disappear. The position of the body is most emphasized by Jehle. According to him the position of the body associated with lordosis of the lumbar spine causes albuminuria which disappears when the lordosis is corrected. Certain distinct conditions, however, must prevail: The lordosis must be arc-shaped and the lowest point of the curvature must be in the region of the first and second lumbar vertebræ. Lordosis in the region of the third and fourth vertebræ does not produce the same effect (Figs. 2a and 2b).

According to Jehle, orthotic albuminuria is lordotic albuminuria. He was able to demonstrate the characteristic shape of lordosis in all his patients with orthotic albuminuria. He further demonstrated that albuminuria could be immediately produced when lying down by artificially inducing lordosis, while albumin did not disappear even in the decubitus in a lordotic child that had been placed in a plaster cast.

Furthermore, Jehle was able to produce albuminuria in healthy persons by the artificial induction of lordosis. As he expresses it: the kidney reacts to this position with a degree of accuracy equal to a physical experiment. So far as children are concerned, there need not even be a predisposition, since every healthy kidney will react as described.

This opinion is contrary to that of the other authors referred to who do not think that the position of the body is the exclusive causative factor, but rather the insufficiently regulated level of the tissue fluids caused by a vasomotor reflex.

The fact that all children will not excrete albumin when brought into the lordotic posture is, according to Jehle, not the consequence of renal resistance, but the difficulty of producing effective lordosis.

Jehle states that the question why lordosis should produce albuminuria is most plausibly answered by assuming that it causes mechanical congestion, and for this reason it is necessary that the lordosis should be located at the level of the juncture of the renal veins and the inferior vena cava, which would produce venous stasis or some other circulatory disturbance of the renal circulation.

Some of Jehle's urinary findings in orthotic albuminuria, such as the presence of casts (often in very large numbers) and red blood corpuscles, are not confirmed in my own experience which has recently been enlarged by a fresh number of examinations. On the other hand, he has also found the characteristic presence of proteid bodies precipitable by acetic acid, and high concentration.

It has already been explained by Senator that congestion of the kidney will lead to the excretion of albumin and casts, and Jehle tries to prove by a number of experiments that this is the exclusive pathogenic factor in orthotic albuminuria which, according to him, is identical with lordotic albuminuria. He obtained in both lordotic and normal children the identical urinary picture which is present in lordotic albuminuria, by compressing the inferior vena cava through the abdominal walls above the juncture of the renal veins; furthermore to eliminate albuminuria in pathological lordosis by preventing in some way a stasis in the region of the inferior vena cava, while albuminuria immediately occurred if both factors, stasis and lordosis, were present.

Jehle continues as follows: "Albuminuria strictly follows mechanical laws and can only be explained on the ground of mechanical causes, and not by reflex processes alone. In the erect posture a stagnation occurs in the inferior vena cava from the kidneys down, which in normal individuals is too slight to cause albuminuria. Nevertheless, there is some slight albuminous excretion after long standing, which would correspond to Senator's 'physiologic albuminuria.' Pathological lordosis increases the stagnation, causing pathological excretion of albumin which is thus merely the sign of exaggerated stasis to which the kidney, as a very sensitive organ, energetically reacts. For this reason artificially and correctly produced lordosis will cause albuminuria in normal individuals. For the same reason albuminuria may be prevented in lordotics by artificially avoiding stagnation. This may, for instance, be done by the upside down position which would prevent stagnation on the ground of hydrodynamic laws, or in the erect position by correcting the lordosis. Finally, it is possible to cause albuminuria in lordosis in the horizontal or even upside-down position, if the slight stagnation which this position involves is increased by a mechanical impediment, such as exists in exaggerated lordosis. Again, albuminuria can be produced in both normal and lordotic children by pressure upon the inferior vena cava above the juncture of the renal veins. Albuminuria is, therefore, induced by causing a mechanical renal stasis, and it is always absent when renal stasis is in some way prevented. Albuminuria is, therefore, only seemingly dependent upon the position of the body, but simply and solely on the conditions of stagnation in the region of the inferior vena cava. Thus, I can induce albuminuria in the decubitus or prevent it in the orthostatic position according to whether I cause or prevent renal stasis by mechanical means."

There is no doubt that Jehle has proved by his ingenious experiments that albuminuria may be caused by lordosis, which has been confirmed by those who have tested his findings, among whom are Bingel, Bruck, Nothmann, Goetzky. Critics should therefore not question the fact of the existence of lordotic albuminuria, but concern themselves with the following questions:—

1. Is every case of orthotic albuminuria lordotic?
2. Does lordosis of the upper part of the lumbar spine always lead to albuminuria?
3. Is Jehle's explanation of the pathogenesis of lordotic albuminuria correct?

The question whether all children with orthotic albuminuria have the characteristic form of lordosis cannot be answered without reserve at the present time, since this would require extensive investigation which at present has only just commenced. I have unquestionably seen cases of orthotic albuminuria without characteristic lordosis and Goetzky's investigations with the large material of the Berlin Children's Clinic have shown that not only are there children with characteristic lordosis and no albuminuria, but also cases of orthotic albuminuria without any trace of lordosis. However, it should be remembered that the clinical examination will only reveal one side of the vertebral column, while we have no knowledge whatever of the behavior of the ventral side, and it is upon this and its relation to the vena cava that Jehle's theory chiefly depends.

As to the consistency of the urine, this may be identical in genuine orthotic albuminuria and in the artificially produced lordotic form, or it may be restricted to the presence of proteid substances that can be precipitated by acetic acid. But the excretion of casts and red blood corpuscles does not belong to the picture of orthotic albuminuria, as has recently again been corroborated by a number of investigations by Goetzky which directly contradict the probability of that affection being present. In Jehle's experiments, however, casts and other form elements were unusually frequent components of the urine in lordotic albuminuria. We would, therefore, have to assume that the lordosis in genuine orthotic albuminuria is so slight as not to cause any material injury to the renal epithelia by whatever process. This, however, would be very remarkable since all degrees of lordosis are represented in orthotic albuminuria. For the present I cannot see any justification for identifying orthotic with lordotic albuminuria and for replacing the name of albuminuria orthotica by albuminuria lordotica, as was proposed by Escherich some time ago.

In regard to the second point, we find that Jehle's experiments themselves and the test cases of other authors have proved that experimental lordosis leads to albuminuria only in a certain percentage of cases. There is no reason whatever to suppose that in the negative cases the spinal cord should offer any particular resistance to the production of an effective lordosis, if we confine ourselves to children and exclude adults. I have seen cases of most pronounced lordosis, in dystrophy, for instance, in which the urine did not contain the slightest trace of albumin. The dictum that the kidney reacts with the regularity of a physical experiment to the position of the body in lordosis, is therefore far from being a demonstrated fact.

The last point concerns the question whether Jehle's assumption is correct that renal stasis by compression of the inferior vena cava explains the pathogenesis of orthotic albuminuria. The fact of its being possible to obtain by pressure on the vena cava through the abdominal walls a urinary picture similar to that in lordotic albuminuria is of course no proof for the identity of both forms of albuminous excretion. In my opinion it is exceedingly difficult to believe that lordosis of the lumbar spine causes compression of the vena cava, as the latter is not a rigid system incapable of evading a pressure. If the pathogenesis of orthotic and lordotic albuminuria were identical and the vena cava were compressed each time upon arising, a short time would suffice to form a collateral circulation which would cause the affection to disappear. Indeed, Jehle himself refers to such a collateral circulation, but only to explain by it the daily vacillations of orthotic albuminuria. He fails to consider that the daily demands made upon it would in a short time develop it to such an extent as to cause the albumin to disappear, which however is not the case. Pfaundler, too, disagrees with Jehle in so far as a causal connection between lordosis and renal insufficiency is concerned. In vain he looks for a plausible explanation in what way the topographic and mechanical conditions can cause compression, tearing, kinking, or any other impairment of the renal blood. Furthermore. Pfaundler points out that Jehle's mechanical theory is at variance with experiments in which albuminuria appeared in an orthotic patient who was immovably fixed and gradually raised to a certain degree from the horizontal position. Increased pressure in the renal veins does not cause albuminuria.

These objections are not intended to detract from the great importance of Jehle's experiments. They are intended to sound a warning not to prematurely derive therefrom the identity of all forms of albuminuria, even the "physiological" form, which in my opinion is still shrouded in complete mystery. Thus, Winternitz unquestionably goes too far in trying to explain the occasional occurrence of albuminous excretion in swimmers, as follows: "Unpracticed swimmers endeavor to keep the head above the water as much as possible, which would of necessity bend the body backward and cause a lordotic curvature of the lumbar spine, especially of the uppermost vertebræ. This would cause a mechanical condition which Jehle considers necessary for the induction of albuminuria. Sponging, douching, etc., do not produce albuminuria because these procedures do not involve a lordotic curvature of the lumbar spine. In fact, when using the bath tub, the upper part of the body is bent forward, and that is perhaps the reason why even after the coldest and longest tub baths traces of albumin are found only quite exceptionally."

In my opinion it would be disastrous for the investigation of the albuminuria question if a uniform opinion were adopted before the foundations upon which the affection rests have been established. All we know is that orthotic albuminuria is a special and peculiar form of a long-lasting renal secretion of albumin which does not depend upon a tissue affection of the renal substance, but is bound up with a certain period of organic development. In all probability it is a functional disturbance in the vasomotor region.

Diagnosis.—"The diagnosis of physiological albuminuria is not particularly difficult, but cannot sometimes be made with absolute certainty, especially if the case has only been observed for a short time." This statement of von Leube may be adopted for the diagnosis of orthotic albuminuria likewise. It is not an instantaneous diagnosis and requires the exclusion of a group of affections with which it has the general conditions in common which have been above described. These, according to von Leube, are terminating nephritis, beginning nephritis, insidious interstitial nephritis, albuminuria of puberty.

I agree with this enumeration, except to express a doubt in regard to albuminuria of puberty. In my experience I have found this to be essentially different to orthotic albuminuria, as there were always the typical method of albuminous secretions and the general conditions associated with chlorosis, without casts in the urine. The affection often disappears as soon as the organism is completely developed, in young women immediately after the first menstruation.

Of much greater importance is the differential diagnosis with nephritis and non-renal affections which are associated with albuminuria, such as cardiac disorders and chronic febrile affections. Cystitis as a cause of albuminuria can be excluded by examination of the sediment, while chronic nephritis may give rise to considerable difficulties, because in this affection the albuminous excretion may present the orthotic type. The investigation of orthotic albuminuria has not yet advanced far enough to admit of a judgment as to whether the chemical composition of the urine may be a factor in the differential diagnosis. Méry's statements in this respect are based upon the urological cycle and are entirely hypothetical. Nor will it be permissible to draw conclusions from the daily quantities of excreted albumin. The method of excretion may possibly be of importance, as will easily be understood from the previous remarks on this subject. Even though alternating excretion of albuminous and non-albuminous urine is an important factor in the diagnosis, it should not be overlooked that some forms of true nephritis may be associated with this symptom. "These forms consequently can not be excluded until an extensive investigation into the presence of nephritis has established a negative result." Hypertrophy of the left ventricle, elevated blood pressure, or increased pulse tension exclude the diagnosis of orthotic albuminuria. Examination of the fundus of the eye should in no case be omitted, since retinitis albuminurica may be an early symptom of nephritis. Headache, vertigo and vomiting may not at once be taken as uremic signs, as these symptoms may also occur in pure orthotic albuminuria. In regard to urinary casts I have already stated my opinion to the effect that their presence would under any circumstances bar the diagnosis of essential albuminuria.

"But if all these findings are in favor of orthotic albuminuria, I want to observe all these cases for at least a year or more, and repeatedly convince myself during that time of the absence of any and every symptom pointing to nephritis, before I make my diagnosis positive." Heubner suggests to facilitate the diagnostic preliminaries by showing the relatives how to make an exact test for albumin with freshly voided urine. The tests should be made four or five times a day for an initial period of one to two weeks, the results being chronicled in tabular form. In doing so, it is important that, before retiring for the night, the child should not urinate into the night vessel, and the same rule holds good for the first morning urine. The urine voided while lying down, should be free from albumin. The total quantity of day urine is collected and examined in all directions, and if there should be any sediment, it should contain no morphological components. These examinations should be continued for a year every two or three months, and during that time the child should receive no treatment whatever, living exactly like a healthy child. Then it will be possible to make a correct diagnosis.

Course.—The course may be distinctly chronic. The condition has been observed for twenty years and longer (Heubner, Posener), as reported in the literature. The albuminous excretion may be interrupted for days, weeks or months (intermittent albuminuria), which will render a decision exceedingly difficult as to whether the albuminuria has come to a definite standstill or not.

Prognosis.—The prognosis of the affection as such is absolutely favorable in the great majority of cases. Heubner holds the same view, although he has reported a few isolated cases in which orthotic albuminuria developed into infantile nephritis after it had persisted for years. Personally I consider the transition of true orthotic albuminuria into nephritis unproven. Probably the cases which are supposed to prove it, were from the first nephritis of an extremely insidious character and had nothing more in common with orthotic albuminuria than the periodicity of albuminous excretion. Von Leube looks upon the form of the affection which may develop into nephritis, as albuminuria of puberty, which he regards as a group of its own. He says: "In cases where cardiac insufficiency and anæmia dominate the pathological picture, parenchymatous nephritis may set in owing to persistent, though light stagnation, and to bad nutrition of the renal parenchyma which may render the epithelia less resistant to renal irritation which under ordinary circumstances would not give rise to nephritis." It is a noteworthy fact that, in individuals with orthotic albuminuria, nephritis does not occur in the course of an infectious disease any more frequently than otherwise; indeed, cases have been reported in which the albuminous excretion was arrested after the infection was cured.

Statistical figures, compiled by Fuerbringer, on the duration and cure of the disease in isolated cases are of great interest. Thus, Langstein had 17 cures in 47 cases within a year; Schur 12 cadets in a short time; Heubner, who furnishes the valuable information that he has convinced himself of the complete health of his patients for a period of nine years, had nine positive and five probable cures. Of Méry's 16 cases 11 recovered; of Schaps' 11 cases 4 recovered. Von Stejskal reports 2 positive cures of 10 orthotic patients and le Noir 1 cure in spite of 3 pregnancies. Kannegiesser re-examined 29 orthotics and found nothing pointing to nephritis in 14 of them. Lommel, Matthes and others take the affection to last from 2 to 2½ years, and this seems to be the rule, although by no means free from exceptions. Neukirch, for instance, reported a cure after the affection had lasted for five years. Finally, cases of 15, 17 and even 20 years' duration have been communicated, some of the patients being more than 30 and 40 years old (Martins, Heubner, Posner, Lenhartz). Thus, orthotics may well obtain a life insurance policy, which no doubt they often do, although the majority contract the disease between the ages of 7 and 15.

Nevertheless, it cannot be denied that there is a certain degree of inferiority in individuals affected with orthotic albuminuria, as evidenced by clinical experience and further illustrated by the fact that these individuals are frequently a prey to tuberculosis. The latter fact, indeed, has led to the suspicion of the existence of close relations between tuberculosis and albuminuria. Furthermore, von Leube cites Washburn's experience, according to which 39 persons with "physiologic albuminuria," whose lives had been insured, showed a mortality of 17.9 as against the expected figure of 9 per mille.

Treatment.—The strict differentiation between orthotic albuminuria and nephritis must needs find adequate expression in the treatment. There would be no sense in ordering children with orthotic albuminuria to abstain from physical exercise. The object of a rest cure in the decubitus is simply to arrest the albuminous excretion for the time being, as it immediately reappears upon the child's rising. It may, however, impair the general condition, appetite and disposition, which may aggravate the complaint. An exclusive milk diet, which is still often prescribed in nephritis, would be complete failure, since it impairs the general condition of the child, increases the pallor of the complexion, without doing any good whatever. Children with orthotic albuminuria should have the same nourishment as healthy children of the same age and go through the same amount of physical exercise. Fresh air living in forest or mountain regions is advisable. So is stimulation of the skin and circulation by massage and friction. Edel advises gymnastic exercises, and they may be tried. It is clear, however, that children should not undergo any excess of physical or mental labor and that they should be protected from catching cold by clothing them in accordance with the weather conditions. In anorexia tonic stimulants are indicated, and iron preparations often do good. In young women with "albuminuria of puberty" the administration of 2 to 3 Blaud's pills 3 times daily has shown excellent results. Should there be simultaneous lordosis I do not—contrary to Jehle—advise the application of a corsage. Even if albuminuria should cease with the correction of the lordosis, it would not improve the general constitution, and that is the object to be worked for. All treatment should be avoided that might impress the child with the fixed idea of being ill, for by doing so, neuropathy might be artificially cultivated, which would be a greater evil than a harmless albuminuria.

HÆMATURIA

Hæmaturia may be defined as the excretion of urine containing blood. Its importance is purely symptomatic, since all conditions of either the excretory or the conducting apparatus, which may be attended by hæmorrhage, manifest themselves by bloody urine. A mistaken diagnosis due to the admixture of blood from some other source than the urinary apparatus may be avoided by thorough investigation.

According to the quantity of the admixture of blood the color may vary from slightly reddish to blood red. Only by microscopic examination can the diagnosis of hæmaturia be definitely established, and the possibility of error due to some other coloring matter be eliminated.

Among the *local causes* of hæmaturia in infancy are to be mentioned, inflammatory diseases of the kidney (especially scarlatinal nephritis), trauma, stone, tuberculosis, tumors, embolic processes, and thrombosis of the renal vein, if it does not produce complete anuria.

Furthermore, we have to remember that the various forms of hæmorrhagic diathesis may be attended by hæmaturia, or even present this as the sole symptom (renal hæmophilia, Senator). This is true of a special disease of infancy, infantile scurvy (Barlow's disease). A number of cases of this disease have been recorded in which the hæmaturia was the only symptom, and in which the bloody urine disappeared promptly under improved diet. It may be remarked that in almost every case of infantile scurvy if the examination is sufficiently exhaustive, blood corpuscles in greater or smaller numbers will be found in the urine (Heubner). Guthrie has described a congenital, hereditary, family form of hæmaturia, which affected twelve members of a family, who were not bleeders. The hæmorrhage appeared in these persons especially after partaking of certain dishes.

Not infrequently it is difficult to determine the source of the blood, and to decide whether it comes from the renal parenchyma or from the urinary tract. In hæmorrhage from the kidneys, unless due to injury of a large vessel, the blood and the urine are intimately mixed, and it is rare to see a coagulum settle to the bottom of the glass. An unerring sign that the hæmorrhage comes from the kidney is the presence of blood-casts (see the cut of sediments from scarlatinal nephritis). But besides these there will be found other casts and renal epithelium. According to Gumprecht, fragmentation of the red blood corpuscles, the finding of numerous microcytes in the urine, is always an indication that the hæmorrhage did not arise below the kidney. He believes that the hæmorrhage is the result of the action of the urea on the blood platelets. According to Heubner, the presence of numerous infinitesimal blood corpuscles in the urine in hæmorrhagic scarlatinal nephritis is due to the impossibility of the larger cellular elements passing through the crural arch.

Just as the **prognosis** in hæmaturia depends upon the fundamental disease, so the **therapy** is governed by the cause. Hæmorrhage from the kidneys demands absolute quiet, a diet free from spices, and the application of the ice-bag. If the hæmorrhage is more persistent, the internal application of gelatin, or the subcutaneous injection of a 2 per cent. solution of gelatin is worth trying. In cases of renal hæmophilia I saw the bleeding arrested by this means. As a last resort extirpation of the bleeding kidney may be attempted. In a case of renal hæmophilia, Israel obtained a cure by peeling out and replacing the kidney.

HÆMOGLOBINURIA

By hæmoglobinuria is meant the discharge of the blood coloring matter in the urine. As a matter of fact, methæmoglobin will be found more often than hæmoglobin, since the latter soon changes to methæmoglobin in urine which is allowed to stand. But the direct discharge of methæmoglobin has been observed (Ehrlich). The blood-pigment is recognized by the spectroscope. The urine may show every shade from a pale reddish tint to a red Burgundy wine color. The urine is albuminous in proportion to the amount of hæmoglobin. In the sediment the red blood corpuscles are either not found at all, or they are in such small numbers that they cannot explain the presence of the hæmoglobin. The blood coloring matter is found frequently in the form of casts or amorphous masses, less often in the form of crystals. Hyaline and granular casts, and crystals of calcium oxalate are seldom absent.

Hæmoglobinuria results from a number of causes. In general it is the effect of a toxæmia, and this must be accepted as the explanation even where we do not know the exact nature of the toxic process. The poisons which cause hæmoglobinuria are well known,—the chlorine salts, phenol, naphtol, sulphuretted hydrogen, toluendiamin, and also the mushroom poison, which has not yet been chemically determined. Passing over the very rare form of congenital hæmoglobinurias the hæmoglobinurias resulting from the infectious diseases (scarlatina, measles, typhoid fever, erysipelas, malaria), and the so-called paroxysmal hæmoglobinuria are of especial interest to the pediatrist.* Among the infectious diseases, aside from malaria, scarlet fever particularly predisposes to hæmoglobinuria. Heubner describes a case appearing upon the twentieth day of the illness with collapse, dyspnœa, and great frequency of the pulse. He attributes the hæmoglobinuria to the action of the same toxins as are responsible for scarlatinal nephritis.

The clinical aspect of paroxysmal hæmoglobinuria merits a detailed description, since it appears occasionally in infancy. By this term is meant the appearance of hæmoglobin or methæmoglobin in the urine in paroxysms, under certain conditions. The most important cause, if not the only cause of this condition is cold. This anomaly is therefore a typical disease of cold.

Symptomatology.—Sooner or later after exposure (cold bath or cold air) the child will complain of malaise, and of chilly sensations. Typical shaking chills have been observed. The temperature may remain normal or it may rise above 40° C. (104° F.). The child soon begins to complain of painful micturition, the urine is of a more or less red color, resulting from the presence of dissolved blood coloring matter. Other symptoms are due to vasomotor disturbance, pallor of the face, slight cyanosis of the lips and ears, cold extremities. After a few hours the child begins to feel better as a rule, the quantity of hæmoglobin diminishes, and after three or four passages of bloody urine the normal quality may be restored. Sometimes the hæmoglobinuria is outlasted by a slight albuminuria. After severe paroxysms icterus may appear and biliary pigment as well as urobilin and urobilinogen may be excreted in the urine. Urobilinuria may be a symptom of abortive cases which do not progress to the escape of blood coloring matter.

Pathogenesis, Character.—A great many theories have been advanced to explain the appearance of hæmoglobinuria. They may be divided into two groups: the one supposes that the separation of the blood coloring matter occurs outside the general vascular system; the other that it takes place within the vessels. Most of those who maintain the first theory consider that the kidney is the organ in which the blood coloring matter is extracted from the stroma of the blood corpuscles. The superabundance of blood in the kidneys which precedes the excretion of the coloring matter is said to be due to abnormal circulatory conditions resulting from cold. Abnormal quality of the blood is said to be a predisposing factor. Another theory asserts that it is in the bladder alone that the coloring matter is extracted by a urine very rich in oxalic acid.

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*The epidemic hæmoglobinuria of the newborn, Winckel's disease, will be spoken of in another place.

Those who defend the theory that the blood coloring matter is separated within the blood vessels, use as an argument in their support the fact that hæmoglobin can be proved to be present during the paroxysms. Some believe that cold is the cause of the blood dissolution; others think it is due to toxic influences, either autointoxication, or infection by toxins from without. Ehrlich believes that under the influence of cold there is produced a substance which will dissolve the blood corpuscles, and that this is autolysin. Chvostek suggested a mechanical destruction of the red blood corpuscles during the paroxysm. In the year 1904, Donath and Landsteiner succeeded by a series of ingenious experiments in advancing our understanding of the character of the affection. They made the following experiment:—During the interval between the paroxysms of a hæmoglobinuric patient, they took the blood, kept it liquid by oxalate of potassium, and preserved it for a considerable time in a cold and in a warm state. Hæmolysis did not appear. But when they cooled the oxalate blood and then placed it in the incubator, the process was followed by hæmolysis. I reviewed this experiment, and proved it to be true. Therefore "the serum of a person with hæmoglobinuria contains a lytic substance which influences the human blood corpuscles. This lysin cannot be demonstrated when the hæmoglobinuric serum is brought directly in contact with its own corpuscles or those from another person, but it may be proved if we consider that its influence depends upon the temperature."

Etiology.—As already stated the immediate cause is cold. Abnormal blood conditions, of which the most important is hereditary syphilis, are predisposing causes. Stempel found syphilis 23 times among 77 carefully investigated cases; *i.e.*, in 29.8 per cent. The same disease was found in Donath's cases in 60 per cent. In three cases of my own, I have proven hereditary syphilis with certainty in two. How far other constitutional states are responsible for the appearance of paroxysmal hæmoglobinuria we cannot definitely determine.

Course.—The paroxysms as a rule last at the utmost only a few hours. In the more serious cases they may be repeated every week, especially during the colder season, while warm weather generally leads to a temporary cure. There are individuals who respond to every exposure throughout their lives by a paroxysm of hæmoglobinuria, but on the other hand cases have been described which recovered after puberty.

The **prognosis** so far as life is concerned is good. Where there is hereditary syphilis, an energetic course of mercury and the iodides may lead to recovery from paroxysmal hæmoglobinuria, at least in some cases. Of the complicating conditions which may lead to a fatal termination, nephritis holds the first place.

Therapeutics.—First in importance from the standpoint of the etiology is vigorous antisyphilitic treatment, even in the cases where the syphilitic connection has not been definitely established. To break the paroxysm itself we have recourse to warmth, hot packs, and hot drinks.

Prophylaxis is naturally of the greatest importance. Individuals subject to paroxysmal hæmoglobinuria should be protected from chilling, and, if their circumstances permit, they should spend the winter in a southern climate.

ANATOMY OF THE KIDNEYS IN CHILDHOOD

Like the suprarenal glands, the kidney in the newborn is greater in proportion to the body weight than in the adult. In the newborn it is as 1 to 82–100, in adults as 1 to 225. The lobulation shown in the fœtus is preserved in the newborn, and may persist for a long time. While the renal hilus is at the level of the first lumbar vertebra in the adult, in the newborn it is opposite the second lumbar vertebra. While in general the kidneys extend from the first to the fourth lumbar vertebra, the limits may vary upwards as high as the twelfth dorsal, and downwards as low as the fifth lumbar vertebra. The index finger introduced into the rectum of the newborn reaches easily to the lower pole of the kidney. Owing to the presence of the liver, the right kidney is pushed down, and lies one half to one cm. lower than the left. In infancy, according to Büdinger, the kidneys are always somewhat movable, both during respiratory movement, and by the finger of the examiner. The older the child the more fixed becomes the attachment of the kidneys under physiological conditions, and beyond the age of infancy they may be regarded as being pretty securely fastened.

EXAMINATION OF THE KIDNEYS, INSPECTION, PERCUSSION, PALPATION

When pathological conditions of the kidneys are suspected it is necessary to examine not only the urine, the importance of which has already been emphasized at the beginning of this chapter, but the body as a whole. The methods of *physical diagnosis* will serve this purpose. They should include not only the examination of the diseased organ itself, but of the rest of the body, especially the heart, vascular system, and the fundus of the eye. Some of the physical methods of examining the kidney in infancy deserve to be employed more often than has been the custom hitherto. These methods are, besides inspection, palpation, and percussion, the use of the X-ray, the cystoscope, the ureteral catheter, and the other methods of obtaining the separate urine from each kidney. It is true, however, that experience with these methods encourages their use only in older children.

The local inspection, according to Strauss, is best made by comparing from behind, one side with the other, first with the child prone, and afterwards in the erect position. In many cases of large renal tumor, or of hydronephrosis, bulging may be detected either at the side, behind, or in the abdomen. The presence of an œdematous swelling or of reddening of the skin near the loins is of value for the diagnosis of inflammatory processes in or near the kidneys, and of peri-, and paranephritic suppuration.

In regard to *percussion* of the kidneys, Steffen says that it is quite possible in any child to determine the percussion limits of the kidneys above, below, and laterally. In diseases of the kidney, if daily examinations are made, and recorded upon the skin with a colored pencil, the variations in volume (increase and decrease) can be distinctly recognized. Other observers believe that the results of renal percussion are of little value, and it is probable that in practice the method is only useful in so far as it controls the results of palpation.

The *palpation* of the kidneys determines the local tenderness to pressure, changes in size, position, consistency, and mobility. It is best made with both hands with the patient in the dorsal position, and it is well to first have the bowels and bladder well emptied. According to Israel, the best attitude in most cases is the semi-lateral, in which the patient lies midway between the dorsal and the full lateral position, so that the frontal plane of the body meets the plane of the table at an angle of 45 to 50 degrees. After the patient has been put in the dorsal or semi-lateral position, place the flattened finger tips immediately beneath the last rib, a little in front of the lateral edge of the longissimus dorsi, and place the whole palm of the other hand, with the fingers fully extended, upon the surface of the abdomen so that the tips of the second and third fingers are beneath the tenth rib. In this manner, according to Israel, by gentle counter pressure from above during deep inspiration, one may even feel small tumor masses of the posterior plane or the convex edge of the kidney glide down from beneath the twelfth rib. In adults, normal sized kidneys which are in their proper location can be felt only under the most favorable conditions, and there are probably only a few physicians who are able to palpate, as Israel does, the lower pole of any normal kidney. In the case of children the conditions are more favorable since the kidneys extend lower towards the pelvis, and this is particularly true in regard to the babe. In babies of less than three months, Knöpfelmacher recommends rectal palpation. "The procedure is as follows:—Place the left hand near the loins, and press lightly with the slightly bent fingers upon the muscles during the introduction of the well greased index finger into the rectum. According to the age, and the size and position of the sigmoid flexure, one will succeed in palpating one or both kidneys." It is generally easier to palpate the right than the left kidney, owing to its lower position on the right side. By this method the lower third or half of the kidney may be explored by the finger. The dorsal or semi-lateral position should be assumed by the child, or if the belly is too rigid, it may be done in the warm bath or under narcosis. While using this method, it is possible to confirm the observation of Büdinger, Israel, Litten, Wolkow, and others that the kidney under physiological conditions, moves with the respiration.

ANOMALIES OF THE KIDNEYS

Anomalies of the kidneys include complete absence of one or both kidneys, disturbances in the frontal development, or else deviations from the normal position (Alsberg).

Absence of the kidneys is generally accompanied by other serious deformities (Zaufal). Hochsinger has described congenital absence of the whole urinary system, combined with total lack of liquor amnii. In this case the suprarenal glands were present, and the sexual organs perfectly normal. A serious deformity of this kind raises the question of how far the kidney is necessary to life during the fœtal stage. Scheib's observation of a female fœtus seven months old, proved that intra-uterine development of a fœtus is possible notwithstanding the absence of both kidneys. The fœtus seen by him lived for ten minutes after delivery in spite of the serious deformity.

Unilateral defect of the kidney, according to Klebs, generally occurs in the left kidney. On the whole, most renal deformities occur on the left side, a fact which this author explains by declaring that the spiral rotation of the body is an important causal factor. According to Ballowitz, one-sided defect of the kidney is often combined with anomalies of the Müllerian ducts, and hyperplasia of the kidney is often accompanied by disturbances of the other parts of the genito-urinary apparatus anatomically essential. Hyperplasia of one kidney is generally conditioned upon hypoplasia of the other. Persistence of the fœtal lobulation, the "reniculi," is not to be regarded as a deformity; it will not infrequently be found even in adults.

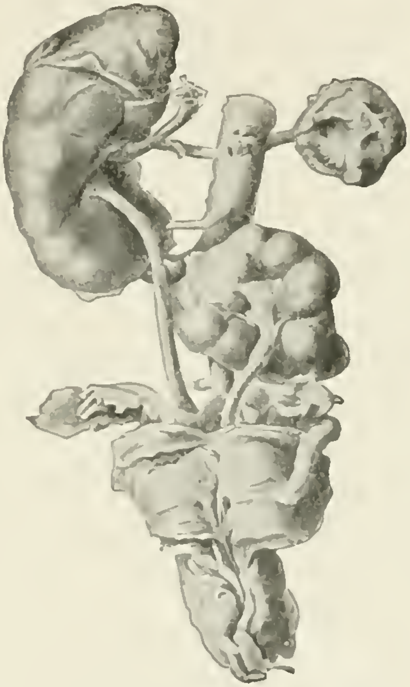
Multi-formation of the kidneys is of the utmost rarity. The finding of two kidneys upon one side has been described by Klebs. Somewhat more frequent is the *horseshoe kidney*, resulting from fusion of the two kidneys either at the upper or the lower pole, or more rarely at the hilum. The horseshoe kidney is as a rule abnormal in location.

Changes in the situation of the kidneys may be either congenital or acquired. Dystopia affects in most cases the left kidney, whose position may vary from the lowest part of the small pelvis to the fourth lumbar vertebra. Lobulation of such congenitally misplaced kidneys is as a rule, strongly accentuated. Contrary to the condition found in acquired dislocations, the ureter is not twisted. The blood supply is often derived from several renal vessels of abnormal origin. According to Zenker, the common dystopia is simply a lower position of the left kindey, with displacement towards the middle line. The direction of the hilum is to the front and upwards. The suprarenal gland is in its usual location.

A case of crossed dystopia of the kidney with change of situation of the sexual organs, has been described by Schumacher.

Dystopia of the kidney is perceptible as a tumor, and the anomaly has some clinical importance owing to the fact that it may be confounded with other tumors of the false pelvis, with undescended testicles with swollen glands, with fecal concretions, etc. Moreover, the differential diagnosis from intussusception and appendicitis has in many cases to be considered. Under these conditions palpation may become of decisive importance to establish the diagnosis. The anomaly may be attended in older children by pains localized near the ilium and radiating into the extremities, or it may cause frequent micturition. In certain circumstances, pressure of the ureter against the ilium may be followed by the formation of a "water-bag kidney" (hydronephrosis, Schott), the practical importance of which has been pointed out by Billroth.

FIG. 3.



Dystopia of the left kidney. Fœtal lobulation.

The formation of a purulent sac in the mislocated kidney has also been described. It is said that this will predispose to fixation of the sigmoid flexure. The assertion that congenitally dislocated kidneys are not movable is not exactly true, since in isolated cases abnormal mobility of dystopic kidneys has been observed (Gruber). But it is true that abnormal mobility is not a sign of congenital dislocation of the kidney, as has been stated by Knöpfelmacher.

FLOATING KIDNEY

The kidney under physiological conditions is slightly movable, so that the distinction between pathological and physiological mobility may not be perfectly distinct. The anomaly may without doubt be congenital, as is proven by the observations of Chamney, Hödlmoser, and Phillipps. (The last was a baby six weeks old, whose kidney was extremely movable.) If congenital, the anomaly is the result of anatomical peculiarities of the mesentery and of the renal blood vessels, and according to Orth, anatomical peculiarities of the mesentery predispose to the development of the acquired form.

Symptoms.—While congenital dislocated kidney is usually situated on the left side, unilateral floating kidney is generally on the right. The reports of Kuttner, Comby, Hollederer, Knöpfelmacher, Blum, and others have shown that floating kidney is nothing like as rare in infancy as was once supposed. Girls are more frequently affected than boys. It may exist without any symptoms, its discovery being made accidentally in the course of an examination for some other condition. There may be a feeling of pressure and heaviness, dyspeptic conditions, attacks of pain, appearing especially where the abdomen is subjected to jar, and radiating into the extremities. However, these symptoms are not pathognomonic of floating kidney alone, since the latter is itself frequently but one phase of a general splanchnoptosis. Those violent paroxysms of pain which are due to torsion of a floating kidney, and which may lead to collapse and fainting, seem to be rare in infancy.

The **diagnosis** of floating kidney is made by palpation. It can be certainly established only when we can detect a movable, smooth, oval or kidney-shaped tumor, easily replaced into its normal position. The differential diagnosis requires ruling out of the same conditions as in dystopia of the kidney.

The **treatment** is local and general. After the kidney has been replaced it must be retained in its normal position by means of suitable bandages. In the more severe cases, nephropexy may be required. Regulation of the intestinal action, and in the case of anæmic, weak individuals, the improvement of the general health by appropriate diet and the use of iron tonics will be of service.

NEPHRITIS

A rational classification of the various types of nephritis should be based upon its etiology (Friedrich Müller,* Ponfick). This method of considering Bright's disease is certainly desirable in infancy, since, because of its dependence upon particular infections and intoxications, frequent at this age, the disease will in most cases exhibit a tolerably characteristic urinary analysis. But it is necessary to emphasize the fact that a complete etiological classification is not yet possible because the etiology of the so-called chronic parenchymatous nephritis is often obscure, its origin cryptogenetic, as in the case in adults.

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* Friedrich Müller says:—"Since the term nephritis can only be translated by "inflammation of the kidneys" this designation should be applied exclusively to a restricted group of renal affections, not sharply limited, and not to those forms which are pre-eminently degenerative in character. In a conception of nephritis are to be included not only the many types of disorder which belong to the group of Bright's diseases, but also the ascending and hæmatogenous suppurations. The term Bright's disease has lost its original significance of a renal disease accompanied by œdema and albuminuria, but a newer and more effective name has not yet been found to replace it. It would therefore be better to abandon the term and to use as a collective noun for the inflammatory as well as the degenerative diseases of the kidneys the name "nephrosis."

It is therefore necessary still to cling, at least in the main part, to the division of chronic diseases which has come down to us from Wagner, and which has been adopted by Heubner.

In reference to the postulate of Ponfick, and in order to provide a classification in which the etiology and clinical anatomy are as closely related as possible, we shall separate the discussion of the nephritis of infants from that of older children. In this way the etiological importance of previous disease will be recognized at least in part. Thus in older children the development of Bright's disease is pre-eminently influenced by the infectious diseases (especially scarlet fever and diphtheria), while in infancy the gastro-intestinal disorders are the predominant causal factor. Recent researches have taught us that the renal affections resulting from congenital syphilis form a group whose pathological anatomy is fairly distinct, and we are entitled to hope that the more perfect we make our examinations and the more carefully we trace the causal factors, the smaller will be the number of cases whose etiology we cannot explain. Meanwhile we are forced to be content with a classification which is perfect neither from the standpoint of etiology nor from that of pathological anatomy.

CLASSIFICATION

I. Nephritis of infancy:

1. Of gastro-intestinal origin.
2. Due to other infections and intoxications, and the so-called primary nephritis.
3. Due to congenital syphilis.
4. Contracted kidney.

II. Nephritis of older children:

1. Acute nephritis
    - (*a*) Scarlatinal
    - (*b*) Diphtheritic
    - (*c*) Due to other infections or intoxications, and of unknown etiology.
2. Chronic nephritis.
    - (*a*) Chronic Bright's Disease (second stage, large white kidney).
    - (*b*) Contracted kidney (granular atrophy).
    - (*c*) Chronic hæmorrhagic nephritis (Wagner).
    - (*d*) Doubtful forms (Heubner).
    - (*e*) Amyloid kidney.
3. Suppurative nephritis.

I. NEPHRITIS OF INFANCY

The course of nephritis in infancy is frequently not attended by any conspicuous symptoms, and it may be only by the examination of the urine, which is often neglected in these little patients because of a fancied difficulty in obtaining it, that the damage to the kidneys is discovered. This fancied difficulty and the neglect of urinary examination is responsible for the fact that the investigation of the etiology, clinical course and pathology of infantile nephritis is still so meagre. Acute nephritis is the most common form, but transition into the more chronic types is also seen. It is of importance in diagnosis to remember that it is not enough to examine the urine for albumin, but always to examine the sediment under the microscope, since in no small proportion of the cases renal casts will be found even where there is no albumin present. And it is certainly true that even a microscopic examination may not prove the absence of disease in the kidneys, because post-mortem dissections have demonstrated serious alterations both in the parenchyma and the interstitial tissue of the kidneys, when during life there had been no sign of pathological changes in the urine. We are especially indebted to Cassel for having pointed out these facts in infancy.

1. *Of Gastro-intestinal Origin*

Kjelberg in 1870, in examining 143 cases of "intestinal catarrh" found albumin, casts, and round cells in the urine in 46.85 per cent. His statements have since been fully confirmed and more carefully analyzed by many writers, amongst whom are Baginsky, Bernhard and Felsenthal, Czerny and Moser, Epstein, Heubner, Hirschsprung, Hoffsten, Parrot, and Widerhofer. Simmonds was the only one who believed that the cause of the renal trouble was not the "gastro-intestinal catarrh" per se, but the suppuration of the renal pelvis which often appears as a complication of the catarrh. Czerny and Moser formerly considered that the uranalysis was the most important clinical expedient to distinguish a gastro-enteritis from a simple dyspepsia, since it was then believed that the latter never led to albuminuria or other pathological changes in the urine.

We know to-day, however, that any disease of the gastro-intestinal tract, whether attended by fever or not, even a simple dyspepsia, may in infancy lead to albuminuria, whose presence alone, it is true, does not necessarily mean a pathological change in the renal structure. This opinion is shared by Czerny and Keller, who emphasize the fact that even the most insignificant dyspepsia in babies a few weeks old and nursed at the breast, may be attended by decided albuminuria.

Among the gastro-intestinal diseases the intestinal catarrh and enteritis of Widerhofer's classification seem to be the principal causes of renal disease. Epstein considers that albuminuria is the most important symptom of cholera infantum. According to him it appears within 24–48 hours after the onset of the diarrhœa, increases with the diminution in the amount of the urine which is peculiar to the disease, and attains its highest point at the acme of the affection.

Symptomatology. During the acute stages the symptoms of renal disorder are obscured by those of the diarrhœal disease, and the former are only discovered by the result of a uranalysis. The urine may contain more or less albumin, casts, renal epithelium, and red and white blood corpuscles. It is only in severe cases that the urine appears red to mere inspection. If death occurs during the acute stage of the gastrointestinal disorder with symptoms referred to the nervous system, convulsions or coma, it may be impossible to say whether or not these symptoms were uræmic. The hydrocephaloid state (Widerhofer), and the serious typhoid condition which may appear after the subsidence of the acute paroxysm, are probably of uræmic origin, just as is the case in the cholera of adults, but it must be admitted that this is not always true of the typhoid symptoms.

In some cases, after the intestinal symptoms have subsided, the only indication that points to nephritis is that the child does not rally, but gives the impression of being still seriously ill, and this is only explained when the urine is examined. In other cases the appearance of œdema points to the kidneys as the seat of trouble. The face becomes gradually pale, waxen and bloated, the œdema spreads to the extremities, and may develop into a general anasarca. The urinary excretion becomes scanty, or there may be complete anuria, and if the urine is passed upon the diaper this may look as if it had been soaked with a blood-stained fluid. Rapid pulse, higher temperature, and slight dyspnœa may complete the picture. Vomiting, convulsions, and somnolence indicate the onset of uræmic intoxication. On palpation, the kidneys often seem to be enlarged and tender to pressure.

The *œdema of infancy* requires a short discussion. At this age it does not always indicate an anatomical renal disease. Although this is sometimes true of adults, it is deserving of emphasis that in babies the idiopathic œdema without the appearance of pathogenic elements in the urine, contrary to the tendency of later life, is relatively a common condition. Wagner drew attention to this fact in 1887, and it has since then been confirmed by the further observations of Hutinel, Cassel, and Stöltzner. This disturbance has been attributed to diseases of the heart, of the vessels, to cachexia, and to toxins. In a case which came to autopsy, Ludwig F. Meyer was able to exclude any anatomical disease of the kidneys by a thorough histological examination. Hutinel has insisted upon the great importance of the ordinary sodium chloride of the food in connection with the appearance of the idiopathic œdema.

The increase in the œdema when the amount of salt is increased corresponds with the decrease which follows the reduction in the chloride. The phosphates and the other nutritive salts have the same influence (Ludwig F. Meyer). We are obliged to admit without reserve the correctness of the theory established by Bartels and others that the appearance and increase of œdema is in proportion to the diminution in the quantity of the urine. This decrease may be either the cause or the consequence of the œdema. Both possibilities must be admitted (F. Müller). According to Stöltzner it is impossible to escape the supposition of a lesion of the capillary walls to explain the œdema. This opinion is strengthened by the beautiful experiments of Heinecke who produced œdema in animals by injecting small quantities of blood serum taken from animals suffering with œdema due to the action of metallic poisons. It is also possible that the œdema may be the result of functional disturbance of the organs which ordinarily take care of the elimination of the salts. This is the function of the epithelium of the urinary tracts. If these epithelial cells are so damaged by a certain percentage of salt in the blood, that they become unfit for their duty, the result is an overcharge of the blood with salt, which will be discharged into the tissues and produce an œdema by carrying the water with it (L. F. Meyer).

Pathological Anatomy and Pathogenesis.—Kjelberg and Hirschsprung describe the pathological changes in the nephritis of gastro-intestinal diseases as follows:—Alterations in the renal parenchyma, high-grade fatty degeneration of the epithelium of the convoluted tubules, turbid swelling of the epithelium of the tubuli recti. Epstein, Czerny and Moser also found alterations in the convoluted tubules:—"the epithelial cells of the tubuli contorti are greatly enlarged, the protoplasm is granular, the nuclei are susceptible of being slightly stained, the Malpighian corpuscles and the tubuli recti are sharply distinguished from the diseased tubuli contorti by an intensive color." In some cases Czerny and Moser found areas of infiltration in the renal cortex, consisting of round cells or of red blood corpuscles, but they never found any diffuse infiltration. Microörganisms were found in the exudate and many of the blood vessels were completely filled with them. In cases complicated by venous thrombosis the relation of this complication to bacterial emboli was established by Czerny and Moser.

The results of the examinations of Czerny and Moser permit us to divide the hæmatogenous nephritis due to gastro-intestinal diseases into two forms: one dependent upon bacterial embolism, the other the result of toxins. A bacteriological examination of the blood will be of service in enabling us to make the differential diagnosis between the two.

Diagnosis.—The diagnosis can be established only by examination of the urine. If we wish to avoid the use of the catheter in children, we may make use of the cone devised by Erlenmeyer, fastened in place by means of adhesive plaster. Catheterization is a method easily practiced, and provided proper asepsis is observed it is free from any danger (Hirschsprung). Englisch has devised a catheter for use in infants with shorter tip and slighter curve.

The **differential diagnosis** from cystitis may be difficult because this condition is often a complication of the gastro-intestinal diseases of infancy (see chapter on cystitis).

Among the clinical symptoms is to be mentioned œdema, which is a valuable diagnostic sign, even though it does not absolutely prove the existence of nephritis. Convulsions, debility, low spirits, vomiting are all too ambiguous to warrant their acceptance as pathognomonic of a uræmic intoxication. But Politzer considers that elevation of the fontanelle, with marked resistence and incompressibility, a valuable sign of nephritis, and mentions also convulsions, sighing and vomiting. With reference to the fundus of the eye the literature is not extensive enough to enable us to draw any definite conclusions.

Course and Prognosis.—The renal symptoms may disappear along with the intestinal disturbance, or they may persist for a little longer. The cases in which the improvement in renal and intestinal symptoms is simultaneous seem to be those which result from toxins. The cases which persist after the intestines are well appear to be due to infection, but this is not true of every case.

Chronic nephritis may develop in any case, but conspicuous symptoms may not be present, and frequently the only diagnostic sign will be found in the examination of the urine. According to Heubner extensive hydrops is especially rare in the chronic cases.

In one of my own cases of nephritis due to enteritis, hydrops persisted for over a year in varying degree, until finally a cure was effected.

It has been definitely proven that in the acute stage, death may occur from uræmia, but owing to the meagre number of clinical and histological reports we cannot determine whether this is very frequent or not. The purely clinical answer to this question is difficult because the very symptoms, which are considered to be uræmic, often admit of another explanation. As has already been stated, the prognosis in the acute cases is not unfavorable, and even in the chronic form recovery may take place even after several years. The prognosis is influenced not only by the severity of the symptoms, but also by the condition of the child's nutrition. The judgment of the child's prospects is decidedly influenced by the proper or improper character of the previous nutriment.

A serious complication, and one which endangers life, is the occurrence of *thrombosis of the veins.* In some cases this accident is manifested by the sudden appearance of blood in the urine, but in other cases it may be followed by complete suppression of the urine. Palpation generally shows marked enlargement of the kidneys. Frühwald has described thrombosis of both renal veins, without previous renal disease, and consecutive infarction of both kidneys, as a complication of cholera infantum. The case was one of typical enteric catarrh, in which a discharge of almost pure blood suddenly appeared. The autopsy showed a marantic thrombosis of both renal veins, with consecutive infarction of the renal tissue. The kidneys were enlarged, to four or five times the normal size.

Therapeutics.—Proper feeding is of the utmost importance. Good breast-milk stands at the head of the list, and if this cannot be obtained pure cow's milk should be given. Where there is repugnance to the milk, it is sometimes more willingly taken raw. Artificial food preparations rich in salt (as Liebig's beef, Kufeke, Theinhardt) should be avoided in order to prevent the tendency to œdema, or to assist in its removal if it is already present. The use of hot packs is to be recommended. The child is put into a bath at 39–40° C. (102.2°–104° F.), remains there for about ten minutes, and is then wrapped in a wet sheet and a woolen blanket in such a manner that the head alone remains free. If there is great weakness a dry linen or woolen covering is used instead of the moist pack. The child remains in the pack for 20 to 30 minutes after the skin begins to act, and perspiration is favored by the free administration of hot teas. The child must be watched very carefully during the process, and any tendency to collapse is met by the administration of camphor subcutaneously. The hot pack should not be used oftener than once a day, because it is not without decided depressing effects. If the pack does not produce sweating, pilocarpine should be given internally (pilocarpine 0.02 Gm. (⅓ gr.) aqua 50 c.c. (1¾ oz.). Give 10 c.c. (2¾ dr.) at the beginning of the pack. In case there is great decrease in the quantity of urine, or if uræmic symptoms evidence themselves, hot poultices should be applied over the region of the kidneys. The sovereign remedy for uræmia is blood letting (see therapeutics of scarlatinal nephritis).

2. *Nephritis due to other Infections and Intoxications, and the so-called Primary Nephritis*

The number of cases of nephritis from the above causes is not as large as those due to gastro-intestinal diseases. It is unusual to meet with scarlatinal nephritis in early infancy, because children of this age are comparatively immune to scarlet fever. We find this affection more often in connection with other exanthemata, such as varicella, and as a complication of pneumonia. Inflammations of the kidneys are also seen resulting from meningitis, erysipelas, angina, pyæmia, tetanus neonatorum, impetigo contagiosa, aphthous stomatitis, and generalized eczema (one of the most severe cases of nephritis which has come under my observation in an infant appeared in connection with a streptococcus catarrh). Heubner dwells upon the frequency of nephritis in infantile scurvy. The nephritis is usually hæmorrhagic in character, and its cure is effected along with the casual condition by suitable dietetic treatment. Infants are more easily attacked by nephritis than are adults in consequence of the external use of toxic substances, such as iodine, tar, styrax, etc., and such application ought therefore to be made only when the condition of the urine can be watched, and the presence of albumin and casts promptly detected.

Besides the forms of nephritis whose etiology is pretty definite, in spite of the fact that neither their clinical nor their pathological history has been perfectly studied, there are others which appear without any definite preceding cause. These are called acute "primary" nephritis. The treatment of these cases corresponds to that which has been advised in the other forms which have already been discussed.

3. *Nephritis due to Congenital Syphilis*

It has long been known that syphilis in adults at any stage may lead to the development of the large white kidney, with œdema and abundant excretion of albumin, and under certain conditions also to the contracted kidney.

The nephritis of hereditary syphilis in infancy has been studied carefully only in the last ten years so as to complete our previous knowledge by accurate clinical observations. Among the writers who have earned special recognition by their work in this connection we may mention Cassel, Hecker, Hochsinger, Karvonen, v. Ströbe, and Störk.

Cassel examined 31 babies with hereditary syphilis between two weeks and seven months of age, and in six of these he found albumin and casts in the urine. Hecker in twelve cases found the urine free from albumin in but two of the children. In all the others he established from the uranalysis the diagnosis of distinct alteration in the kidneys. There were six cases with marked parenchymatous nephritis, characterized by abundant albumin and casts while in four there were traces of albumin. Karvonen found a pathological change in the urine but once in six syphilitic infants, of whom two were premature. We have not as yet sufficient material to draw any far-reaching conclusions, but at any rate it is striking that the majority of examiners found clinical evidences of nephritis in only a small percentage of their cases, while they all lay stress upon the pathological histological alterations in the kidneys of both mature and premature syphilitic infants.

The pathological changes in the excretory apparatus of the kidneys consist in more or less serious parenchymatous degeneration. The epithelium of the tubuli contorti as well as of the glomeruli may become affected. Cystic degeneration of the glomeruli and the urinary ducts is often found. The damage done by the syphilitic virus is characterized by pathological alterations in the connective tissue and the vascular apparatus, especially peri-adventitial deposits. To this there may be added deficient development of the glomeruli and of the tubuli uriniferi, so that their number may be considerably less than in the normal kidneys.

The nephritis of hereditary syphilis bears no relation to the severity of the fundamental disease, and the diagnosis is therefore correspondingly difficult, because, on the one hand the uranalysis may be absolutely negative, notwithstanding the fact that the pathological alterations in the kidneys may be well marked, and, on the other hand, when the urinary reaction is positive, it is by no means easy to exclude other etiological factors, especially the influence of mercury in cases which have received specific treatment. Conspicuously large quantities of albumin such as are seen in the syphilitic nephritis of adults, do not appear to be the rule in that of infants; renal hæmorrhage and œdema have been but seldom observed (Finkelstein); and uræmic symptoms in this affection have not been observed.

The paucity of the literature prevents our answering the question whether a complicating nephritis affects to any great extent the prognosis of an individual case of hereditary syphilis. So much is certain; with the subsidence of the general symptoms the pathological alterations in the urine may also disappear. But it is equally possible that there may develop a typical contracted kidney, with plentiful clear urine, of low specific gravity, small percentage of albumin, and very few casts.

Therapeutic measures are directed to the cause. Especially to be recommended are gluteal injections of corrosive sublimate, as advised by Immerwol 0.2 c.c. (3 drops) of a solution of sublimate 0.1 Gm. (1½ gr.), sodium chloride 0.2 Gm. (3 gr.), aquæ destill. 10. c.c. (2¾ dr.) given once a week. This treatment is to be interrupted only if the symptoms of renal disease are increased under the medication. The child is to be fed on the same principles as any patient with congenital syphilis.

4. *Contracted Kidney*

Granular atrophy occurs congenitally in a very few cases (Arnold, Westphal, Baginsky). The transition in infancy from an acute nephritis to the contracting kidney is not very frequent, but it has been shown to occur in the case of the nephritis of syphilis. Heredity certainly has a decided influence in the causation of contracted kidney in infancy. This was especially noticeable in the observation of Hellendall upon a brother and sister of one half and two years respectively, who died of contracted kidney. The mother also suffered from the same condition, and it was shown that the origin of the trouble in the latter coincided with the fœtal life of the children.

The **clinical manifestations** are often very insignificant. The quantity of urine is large and the specific gravity is low. Repeated examinations may discover albumin and a few casts (hyaline), but since they are frequently absent for considerable periods, the differential diagnosis from diabetes insipidus may be difficult. Cardiac hypertrophy and accentuation of the second aortic sound sometimes assist in the diagnosis.

The **course** may be exceedingly chronic. A secondary contracting kidney is always a point of lowered resistance from which, if its powers are taxed, there may proceed symptoms of insufficiency. *Therapeutics* are of little avail. Even a rigorous milk diet continued beyond the period of infancy must be considered injurious in nephritis.

II. THE NEPHRITIS OF OLDER CHILDREN

1. *Acute Nephritis (Acute Bright's Disease)*

Acute Bright's Disease is defined by Wagner as follows:—"A renal disease in which the urine will be scanty for days and weeks, in which albumin is found, and in which the different forms of casts are present in varying quantities, and in which there occur white or red blood corpuscles, and epithelium. Besides this there is sometimes pain in the region of the kidneys, and often frequent micturition. There is a varying degree of general disturbance and in serious cases, after a few days or weeks, there is added dropsy of different organs, uræmia, and inflammation. After a course varying from a few days to several weeks, either a complete cure is effected or the disease becomes subacute or chronic, or else death results. It is rather unusual to be unable to assign a cause; in most cases the cause of the illness is some serious infectious disorder. In a certain number of cases the acute Bright's disease is the only thing that can be demonstrated, but much more frequently there exists some other disease, either still at its height (many of the acute infectious diseases, such as diphtheria, typhoid, pneumonia, acute or chronic external or internal diseases), or else convalescing or completely terminated (scarlet fever). The symptoms of acute Bright's disease are therefore frequently mixed. Some, which proceed from the renal disease, are indicated by the urine; others are due to the original diseases. It is sometimes very difficult to decide whether the latter (*e.g.*, fever, cerebral, and gastric symptoms) are due to the renal or to the primary affection."

Postinfective nephritis probably does not, in the majority of cases, result from the direct action of the bacteria, but from toxic influences. Many bacteria pass in the blood stream through the kidneys (staphylococci, streptococci, typhoid, coli communis, and tubercle bacilli), but this does not necessarily lead to inflammation or abscess formation.

The prototype of the acute infective nephritis which is due to toxic influences is the scarlatinal nephritis, of which Wagner has distinguished two forms: the initial and the ordinary variety. It seems preferable, however, to reserve the term scarlatinal nephritis for that renal disease which exhibits special clinical features and histological symptoms, and which appears only after the entire remaining process seems to have terminated. Albuminuria, with casts occurring at the acme of the eruptive disease would be better designated as "albuminuria scarlatinosa." This affection evidently belongs to that great group of albuminurias which have so intimate a relation with the fever that notice of the connection has been shown by the denomination "febrile albuminuria." With good reason Lüthje has emphasized the fact that in this form of albumin secretion also there are probably inflammatory and degenerative changes in the renal structure, but it seems proper, nevertheless, to restrict this variety of albuminuria with casts, because it commonly disappears entirely with the subsidence of the fever, and because its anatomical basis is not yet perfectly understood. Another renal affection, the "septic nephritis," which accompanies scarlet fever, must be separated from the scarlatinal nephritis because in a strict sense it has nothing to do with the scarlatinal process proper. It is not distinguished in any way from the nephritis which accompanies other septic conditions, and with which its symptomatology is identical.

(*a*) Nephritis Accompanying Scarlet Fever (Nephritis Scarlatinosa)

This affection is a relatively frequent complication of scarlet fever. Heubner in a series of 358 cases observed 36 cases of nephritis scarlatinosa, *i.e.*, almost 10 per cent.; in another series of 393 cases he observed the characteristic renal disease 77 times, *i.e.*, in 19.6 per cent.

The **underlying cause** of the nephritis scarlatinosa seems to be different from that which produces the exanthem and the acute reaction. Neither poison has as yet been isolated, and we know only one factor in the whole process which predisposes to the appearance of this dreaded complication, and this is the "epidemic tendency," a factor which was recognized by Wagner. We now know that there are epidemics in which nephritis appears in scarcely 5 per cent. of the cases, and others in which it occurs in more than 70 per cent. (Steiner, Johannesen).

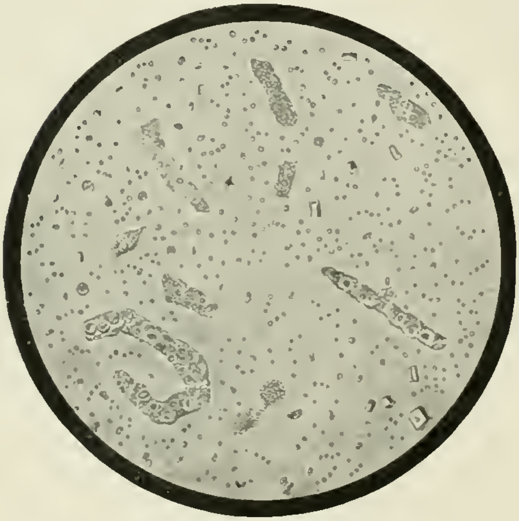
Nephritis does not appear to have any special predilection for the cases which are severe from the beginning. Heubner, out of his rich experience, confirms the fact that the abortive cases, with slight constitutional disturbance, and in which the exanthem was almost overlooked, are at least as often followed by the renal disease as are the severe types, and we are as little able to avoid the complication by any therapeutic measures, as we are able to infer the probable future kidney complication by any symptom occurring in the beginning of the disease. The influence of cold, frequently assigned as an indirect cause, may be denied, since nephritis scarlatinosa probably affects those whose nursing was above criticism as it does patients who go through the scarlatinal attack under the most unfavorable hygienic conditions, with deficient nutrition, and without any proper regulation of the diet. We may consider that the statement made by some writers has been refuted, that in cases in which nephritis appears the disorder was present from the beginning, but was unrecognized because its symptoms were masked by those of the general disease. On the contrary, the examinations of Thomas and Heubner have certainly proved that after the stage of albuminuria the urine will return for a time to the normal before a nephritis scarlatinosa suddenly appears. Furthermore the alterations in the kidneys are minimal in persons who die in the first week of scarlet fever. The observations of Rosenstein and Lenz, to the effect that scarlatinal nephritis occurs most frequently during the stage of desquamation at the end of the third week, have been confirmed. Among 36 cases which Heubner followed from the onset, the nephritis began 17 times between the twelfth and fifteenth days; 10 times between the seventeenth and nineteenth days. The renal inflammation appears therefore at a time when the rest of the disease seems to be over. In the widest sense of the word it is a late effect of the infection. Heubner says that perhaps a special organic susceptibility or lack of resistance influences the appearance of the renal inflammation. The new experiments in etiology have sufficiently demonstrated the occurrence of such a relation between quite isolated cell groups and certain poisons, especially poisons of the group of parasitic toxins or products of dissolution. The family disposition to scarlatinal nephritis which has often been observed, is of interest. It is certainly impossible to determine whether the conception of the kidneys as the point of least resistence is well founded.

Symptomatology.—The appearance of nephritis is often shown by no other symptom than alterations in the urine, which are generally characteristic, and which should therefore be promptly investigated.

The first albumin may be shown in a light, almost clear urine (Burger). The sediment in these cases is scanty, and consists only of isolated casts, red and white blood corpuscles, and epithelium. The corollary of this onset is the form in which casts are secreted without albumin, which shows the importance of both chemical and microscopic examination at the time when we fear the appearance of a renal complication. Gradually after some days the urine may take on its characteristic appearance and then the attention of even the layman may be attracted to it. This may be present from the beginning. The urine is dark, turbid, and of varying shades of reddish brown. The quantity diminishes in proportion to the depth of the color, and the specific gravity is at first increased (1.025–1.040), but later on it falls to 1.015

and lower. The albumin which is always present at this stage may be so plentiful that the urine when boiled coagulates in the tube in a solid mass (1 per cent. and more). The coagulated albumin is colored a dirty brown by the adherent blood-pigment. Above all the sediment is characterized by the conspicuously large number of red blood corpuscles, which are a sign of the hæmorrhagic character of the scarlatinal nephritis. Besides hæmoglobin containing blood corpuscles, extraordinarily small ones are to be seen in addition to the normal sized ones. The presence in the sediment in the form of a brown detritus, of fragments of blood coloring matter, is an indication of the dissolution of the red corpuscles, which may take place in the kidneys themselves. Casts may be present in great numbers—hyaline of every size and form, blood casts, and epithelial casts. Renal epithelium, leucocytes and fatty cells are all far less numerous than the red blood corpuscles.

Fig. 4.



Sediment of the urine in a case of scarlatinal nephritis. In the sediment predominance of red blood corpuscles.

The daily examination of the urine for albumin and formed elements is as important for the diagnosis of the beginning of the renal disease as is the continual estimation of the daily quantity. For neither the daily quantity of urine, nor the percentage of albumin nor the quality of the sediment, inform us of the approaching danger. The less the quantity, the greater, as a rule, the danger of the development of uræmic symptoms.

V. Pirquet considers that the weight curve is of diagnostic value in the beginning of acute nephritis, because the retention of water occurs in most cases earlier than the albuminuria. According to him, the renal inflammation generally goes along with an increase in the weight which results from a retention of water, and this increase shows a typical curve on the weight chart. He believes this increase in weight of greater prognostic importance than the detection of albumin.

As to the other clinical symptoms the *onset* may be quite different. In some cases there may be scarcely any systemic disturbance; in others there may be present a distinct feeling of sickness, with restless sleep, headache, vomiting, and anorexia. The pulse and temperature may be elevated, the latter as high as 40–41° C. (104–105.8° F.), with chill and subsequent sweating, and the fever may continue for days or even for weeks. Slow pulse has been observed by Heubner despite the temperature. Pallor and swelling of the face are in most cases a conspicuous initial symptom. The pallor is the result of an abnormal distribution of the blood, and perhaps also to the decrease in the percentage of hæmoglobin at the beginning and during the course of the nephritis, which has been studied more exactly by Widowitz. According to Heubner an attentive examination at the beginning will almost always reveal œdematous swelling of the cutaneous tissue over the sternum and the anterior surface of the tibia.* A painful swelling of the bronchial lymph-nodes appearing simultaneously with the onset of the nephritis, is often conspicuous.

During the later stages cases differ widely in their *course.* In the mild cases the condition of the urine which has just been described, in which the quantity will hardly sink lower than 400 c.c. (13½ oz.) lasts without much aggravation for one to two weeks.

Gradually the urine becomes clearer and more plentiful. The albumin and sediment noticeably disappear, but not simultaneously, since the presence of sediment may still be demonstrable after the most delicate tests for albumin fail to produce any precipitation.

In serious cases the symptoms are increased and new ones are added to the picture. The kidneys are enlarged and painful to pressure, while the spontaneous pain, generally located in the abdominal region, is comparatively slight. The attacks of colic described by Israel have never been observed by Heubner.

The *dropsy* increases and gives the patient, who is generally extremely pale, a characteristic appearance. It is at first limited to the face, particularly the eyelids, but by degrees it spreads over the extremities, the genitals, and in the end becomes a general anasarca. Serious cases almost always show dropsy of the cavities, ascites, hydrothorax, hydropericardium. Œdema of the mucous membranes may be absent where the œdema of the skin is intense, but may be present if this should be moderate.

The *effect of the œdema* depends upon its location: œdema of the skin causes painful tension and interferes with free movement; accumulation of water in the serous sac gives rise to dyspnœa, cough, and cardiac weakness. Œdema of the glottis may be a complication which seriously threatens life. A high grade of dropsy, without albuminuria, has been observed not very rarely after cases of scarlet fever (Hamilton, Bartels, Henoch). In the cases of Bartels and Henoch the amount of urine passed was extraordinarily small. We must not forget that in addition to the renal and vascular changes, a third component may contribute to the development of œdema in the course of nephritis scarlatinosa. This is an extraordinarily early involvement of the heart. We may say almost without reservation that a clinical and probably an anatomical change in the heart is demonstrable first of all in scarlatinal nephritis, more frequently than in any other form of acute Bright's disease. This was well known to both Bamberger and Wagner but Friedlander studied it more carefully. He found dilatation, generally of the left ventricle, either alone or combined with hypertrophy. The weight of the heart exceeded the normal average by nearly 40 per cent. Silbermann often observed that the changes in the heart began even in the first week. In addition to œdema, the clinical symptoms of cardiac insufficiency are small and frequent pulse, precordial pain, severe dyspnœa, which renders the condition very distressing.

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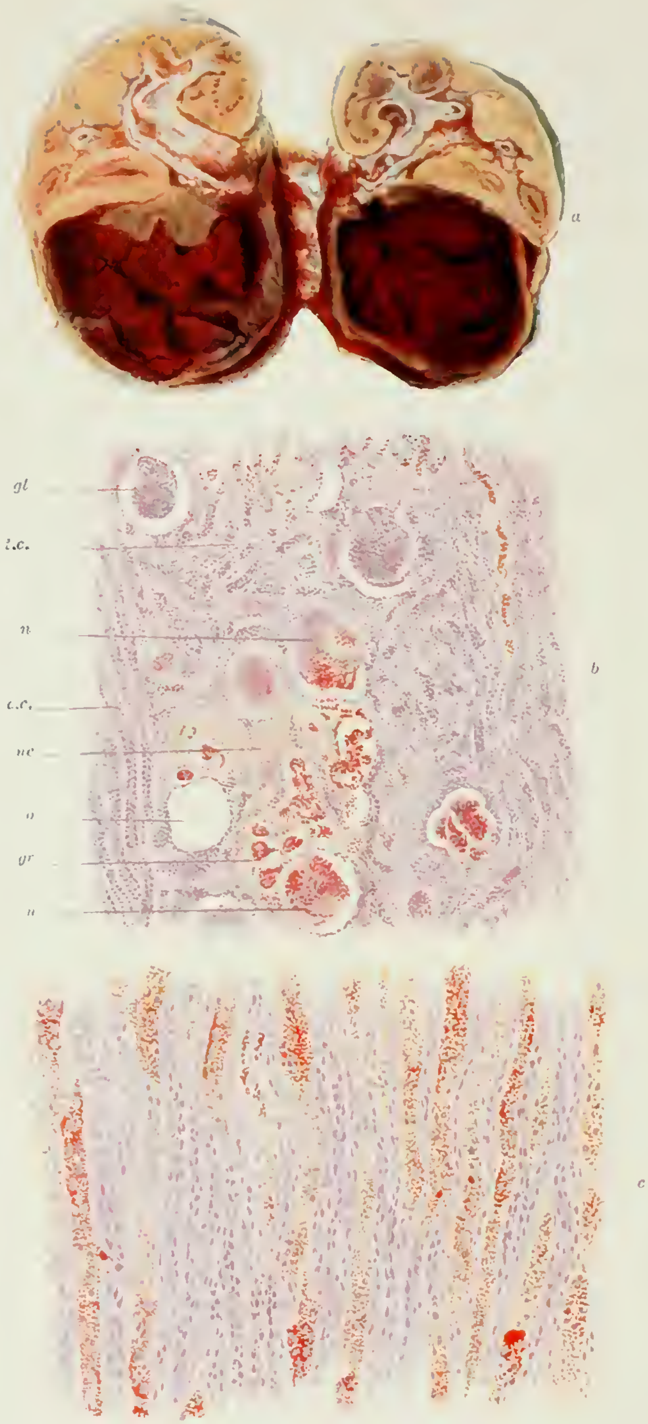
* Bartenstein described a hyperæsthetic (Head's) zone in nephritis from the tenth to the eleventh dorsal vertebral level, between the anterior and posterior axillary lines, situated between the thorax and the iliac crest.

Among the prominent symptoms of nephritis the physician has to give special attention to *uræmic symptoms.* These are generally ushered in by a conspicuous decrease in the quantity of urine, to 200, 100 c.c. (7–3½ oz.) or less. Even complete suppression is not rare, but there are exceptions to this rule, in which the dreaded syndrome appears without any decrease in the quantity of urine. Headache, vomiting, jactitations, increased restlessness and nervousness in the child, serious lack of appetite, the appearance of a thick fur on the tongue, ammoniacal fetor of the breath, should prepare us for the appearance of eclamptic paroxysms. Occasionally, in the midst of apparent good health, the dangerous condition arises without these forerunners. The uræmic convulsions are epileptiform in character; sometimes restricted to single groups; sometimes involving the whole body. They may repeat themselves, or they may be repeated at short intervals, during which the patient either lies deeply comatose, or else exhibits the greatest restlessness or the most violent excitement. During this period the child may die suddenly, without regaining consciousness, or there may be only one or two convulsions, which are quickly followed by recovery. The secretion of urine is restored, and the urine, at first deeply tinged with blood, gradually becomes paler, and within a few weeks the renal inflammation is entirely healed.

It is true that the convulsive stage may be followed by a period which is complicated by sundry disturbances of the central nervous system. The parents are frightened by disturbances of the organs of special sense, of which the most frequent probably is temporary blindness, uræmic amblyopia and amaurosis. These may be the only symptoms of uræmic intoxication, and may appear without premonitory symptoms. The ophthalmic examination is negative, and therefore the cause is probably a toxic functional disturbance in the cerebral centres, which disappears again in a few hours or days. Less frequent are posturæmic deafness, aphasia and paralysis restricted to isolated nerve centres; psychoses may also result from the uræmic intoxica-

PLATE 56.

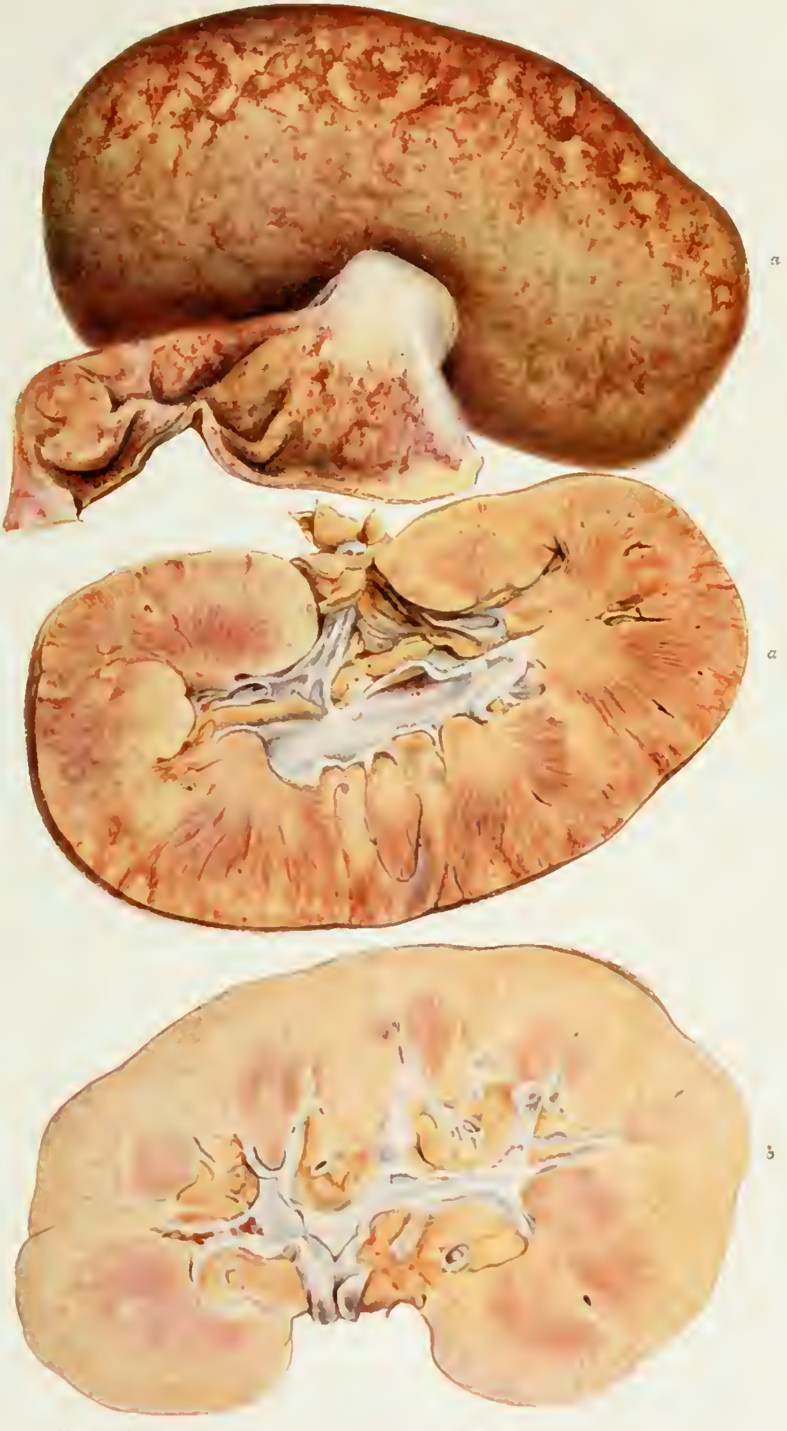


a. Hæmorrhage into the adrenal gland.

b. Acute nephritis in scarlet fever. Child died after 9 days' anuria. *gl*, glomeruli; *t.c.*, tubuli contorti; *n*, necrotic glomeruli with hæmorrhage; *c.c.*, cortical columns; *nc*, necrotic tubuli contorti; *o*, opening due to falling away of glomerulus; *gr*, group of injected capillaries.

c. Acute nephritis in diphtheria. Section through the pyramid.

PLATE 57.



a. Hæmorrhagic parenchymatous nephritis in scarlet fever.
b. Large white kidney. Recurring parenchymatous nephritis with fatty infiltration.

tion, but this may just as easily appear independently in the course of scarlet fever.

In some cases these symptoms of acute uræmia may not appear at all; the affection taking a more chronic course. Sometimes it will present itself in the guise of vomiting and diarrhœa; sometimes in the form of singultus and asthma. Certain patients suffer from violent itching of the skin; others show nervous symptoms, such as headache, vertigo, sleeplessness; all of these forms are of the greatest importance because they have a decisive influence upon the therapeutic measures which we must employ.

In proportion to the multiplicity of uræmic symptoms we may infer a variety of uræmic poisons (F. Müller). Uræmia is certainly connected with the retention of toxins, which are still unknown, but for the secretion of which the kidney has become insufficient. Ascoli connects uræmia with the entrance into the organism of small fragments of renal cells, and with the formation of antibodies, the so-called nephrolysins; but his opinion is absolutely hypothetical.

There are still to be mentioned the inflammatory complications of various organs which appear in connection with nephritis, and which change a fever-free course into a feverish condition, or increase an existing fever. These complications damage not only the organs of respiration (bronchitis, pleuritis, pneumonia), but also the heart, on whose peri and endocardium there are in many cases localized inflammation.

Pathological Anatomy.—The hæmorrhagic character of scarlatinal nephritis is exhibited in its pathological anatomy, in children who die in the earlier stages of the disease. The kidney will be found in a condition of hyperæmia; if death results from uræmic complications the hæmorrhagic changes are more or less marked. The accompanying illustrations (Plates 56 and 57) distinctly show the characteristic renal alterations. The first shows the kidney of a five-year-old boy who died in the middle of the third week as a result of a most serious nephritis with uræmia. The kidney shows an appearance which reminds us of certain many colored marbles; grayish white areas are limited by hæmorrhagic zones of a deep red color, microscopically the inflammation corresponds to a hæmorrhagic glomerulitis, the capsular spaces are filled by hæmorrhages from the hyperæmic capillary loops, just as are the tubuli near the glomeruli whose epithelium, together with the blood vessels, hardens into a coagulated mass. The necrosis of the glomeruli is often quite extensive, where the epithelium is preserved it shows damage to its parenchyma, and is filled with larger or smaller fat-drops. While according to Heubner the loop portions are only rarely the seat of hæmorrhagic infarctions, hæmorrhages will be found in the intercalary parts of the renal ducts. The collecting tubes may to a great extent be obstructed by hyaline casts, a condition which leads to the retention of urine in the adjoining parts. The character of the nephritis which results from the scarlatinal poison, is therefore primarily a disorder of the vessels which is most distinctly in the glomeruli, and has given to the scarlatinal nephritis the name of glomerulonephritis. After the affection has lasted a fairly long time, the aspect of the acute inflammation will change, later on, it will show interstitial cell accumulation with increase of the connective tissue, and subsequent contraction.

Diagnosis.—If it is possible to follow the development of the renal inflammation in the course of scarlet fever, its diagnosis may be made with ease and certainty. If the exanthem has not been observed, the peeling which may still be present, may lead to a diagnosis, but even without this the knowledge that persons in close contact with the child have suffered with scarlet fever within a short time, gives us the right to make the diagnosis, the possibility of which should be taken into consideration in every case of hæmorrhagic nephritis beginning acutely. Still, under these circumstances it is hardly possible to establish more than a probable diagnosis, since in infancy the nephritis which attends other infections and intoxications, may likewise be hæmorrhagic.

Course and Prognosis.—In slight cases without severe hydrops or uræmic symptoms, a cure is usually effected in the course of three weeks. In serious cases, in spite of high grade œdema and uræmic intoxication, it is still possible to obtain recovery in the course of weeks or months. The marantic condition which develops in connection with a nephritis as a result of heart insufficiency, often becomes a special danger. On the whole, it may be said that the more serious the associated general conditions, the more dangerous is the nephritis. Persistent high or remittent fever, tachycardia and brachycardia, lack of appetite and serious intestinal disturbances, aggravate the prognosis. The same may be said of anuria, which almost always terminates fatally if it persists for several days. Family tendencies and the vicarious power of the skin must be considered in establishing the prognosis. In spite of the dangers involved in these statements, the prognosis of scarlatinal nephritis is relatively favorable. According to Heubner six-sevenths of the cases are cured. Whether scarlatinal nephritis often becomes chronic or not, has not yet been unequivocally answered. Unfortunately, there is not yet at our disposal a sufficient number of lengthy observations to determine this question; a factor which is of particular importance in judging of the nephritis of babies. Heubner believes that chronic nephritis is not so rare as a sequel of the acute form, but that the latter will seldom be diagnosed, because the objective disturbances are, as a rule, not sufficient to induce us to examine the urine. The pallor of the child is the only conspicuous symptom; sometimes they complain of headache. The uranalysis in the chronic stage shows a greater or lesser percentage of albumin, and casts in greater quantity, but this is only true during acute exacerbation, which may present the appearance of the original acute inflammation, with all its complications. As long as red blood corpuscles continue to be present in the urine, the chances of an ultimate cure are always greater than when they are completely absent, and when the albumin coagulated by boiling shows a pure white tint. Heubner and Dixon Mann have described a contracting kidney secondary to scarlatinal nephritis, which is incompatible with long life.

To the process of recovery from acute nephritis, the night urine first becomes free from albumin, while the urine passed during the day still contains it. It is advisable to differentiate this form of orthotic albuminuria from the so-called essential one. In this form a thorough examination will almost always reveal the presence of casts.

Treatment.—The question as to whether there is an effective dietetic prophylactic treatment for nephritis, may be answered in the negative in the very beginning. Even though there be no reason to desist from a milk diet during the first weeks of scarlet fever, if the children take this food willingly, it is wrong to force its use if they rebel against it. Cereal gruels and a vegetable diet do quite as well, and are not more prone to be followed by nephritis. Widowitz asserted that by giving urotropin during the first days of scarlet fever, and at the beginning of the third week the development of scarlatinal nephritis might be prevented, but his assertion has been refuted, not only by Garlipp, but by numerous cases in Heubner's Clinic. Weigert believes that since urotropin is harmless, its application may be recommended until further experience in every case of scarlet fever.

Heubner recommends while the renal disease is active, a plentiful supply of milk, which is of value as a mild diuretic. Vegetable diet and cereal gruels may be given without hesitation (von Noorden). The time is past when it was considered a serious fault to replace milk by any other kind of diet. A meat diet is to be avoided, because Weigert demonstrated by examination of the metabolism that it has a most unfavorable influence upon the excretion of albumin. It will be advisable to encourage diuresis by a liberal supply of some mineral water (Wildunger, Biliner).

Special therapeutic measures must be taken to meet a marked decrease in the amount of urine, the beginning of uræmia, and the appearance of high-grade œdema. If the amount of urine falls below 500 c.c., we must resort to hot baths, 35°C. (95° F.), gradually increased to 38° C. (100° F.), in which the child is kept for from 10 to 15 minutes, after which it is wrapped in dry or wet cloths. A liberal supply of tea, as well as the previous internal use of pilocarpine (see the treatment of infantile nephritis), furthers sweating. Hot poultices over the region of the kidney (three times a day for two hours), are frequently of equal value to produce diuresis. Should the quantity of urine sink still lower or serious symptoms of uræmia appear, there should be no hesitation in performing venesection. About one-tenth part of the blood should be removed (the total quantity of the blood is about one-thirteenth of the body weight), either by venesection or by the application of leeches in the region of the kidney (an active leech removes about 10 Gm. (2½ dr.) of blood; as much as will leave the body in bleeding afterwards). The withdrawal of blood may be followed by an infusion of common salt. A convulsion may be effectively combated by a large dose of chloral hydrate, 1 to 2 Gm. (15–30 grains), introduced by enema.

Heubner warns us of the possibility that effective diuretic medicine may increase the hyperæmia. In case cardiac weakness develops, we use first of all injections of camphor, 0.05–0.1 Gm. (¾–1½ grains) two or three times in an hour). Heubner recommends digitalis only in cases of desperate heart weakness. According to Steffen the secale cornutum (extr. fl. secal. cornuti 3–4 Gm. (45–60 grains), aqua 100 c.c. (3½ oz.), 10 c.c. (2½ dr.) 3 to 4 times daily), is of value in the weakness resulting from cardiac dilatation.

If there is marked œdema, a diet containing very little common salt is of the greatest importance (Strauss). Weigert in common with other writers has shown that a salt free diet without any other medicine, may cause the complete disappearance of œdema. If the accumulation of water depends not so much upon the insufficiency of the kidney as upon that of the heart, we need not refrain from the use of stronger diuretics (caffeine, diuretin).

After the subsidence of the clinical symptoms of nephritis, the patient may be allowed to leave the bed for a few hours during the day, but he must abstain from any bodily exercise. Whenever albumin reappears in appreciable quantities, and red blood corpuscles are found, strict recumbency must be again insisted upon.

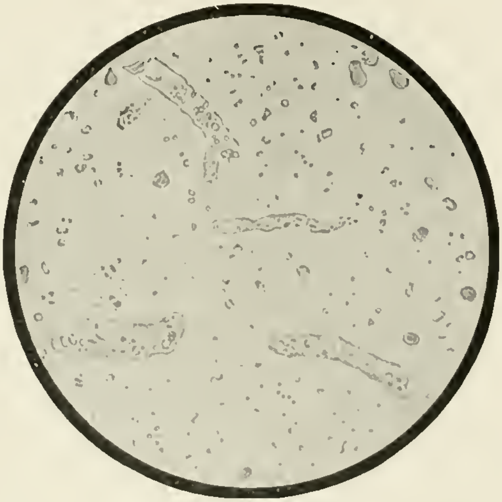
(*b*) Nephritis Due to Diphtheria

There occurs frequently in diphtheria a renal disease which clinically, as well as pathologically, exhibits an entirely different aspect from that of scarlatinal nephritis. According to several statistical tables, symptoms of nephritis are found in from 15 to 65 per cent. of all cases of diphtheria. This complication, according to Unruh, is an absolutely certain sign of a general infection. The renal damage which follows diphtheria is just as little the result of the direct influence of the bacteria as is the nephritis of scarlet fever, but the cause is in both cases to be found in the influence of a toxic action.

Symptomatology.—The urine is normal in color. The specific gravity is increased, the percentage of albumin is moderate, and there is a more or less plentiful sediment consisting of hyaline and epithelial cylinders, leucocytes and fat-grain cells. The number of red blood corpuscles is relatively smaller than in scarlatinal nephritis (Fig. 5). Wagner found a larger percentage of blood only in the most serious cases, with gangrenous changes in the nasopharynx. The amount of urine is seldom less than 200 c.c. (6¾ oz.) and anuria of long duration was not observed. The slight tendency to œdema, uræmia, and to severe general symptoms has been emphasized by all observers.

Pathological Anatomy.—The most important lesion is found in the parenchyma. Heubner lays stress upon the fact that the convoluted tubules and the descending limbs of the loops of Henle, are usually most affected, while the ascending limbs and the intercalary parts are very slightly altered. The collecting tubules also show very early a shedding of epithelium and obstruction by hyaline casts beginning near the cortex. The changes in the epithelium consist in the accumulation of fat drops (see Plates 49–50). In serious cases the epithelium loses its nucleus and shows an indurated appearance. If we remember the location of the most serious alterations, it is evident that we cannot get a clear idea of them by examining the sediment in the urine, because owing to the closeness of the passage through the ascending limbs of the loops, casts and epithelium from this region cannot escape into the urine (Heubner). The fundamental difference in the influence of the scarlatinal and the diphtheritic poison upon the kidney is explained by the pathological anatomy. While the scarlatinal toxin has the greatest affinity for the vasa, the diphtheritic toxin does not touch them, but only attacks the parenchyma. Experiments also teach us the affinity of certain toxins for certain portions of the kidney. Paul Ehrlich, for instance, demonstrated that vinylamin causes a complete necrosis of the medulla, *i.e.*, of that part which represents an evolutionary unit.

FIG. 5.



Urine sediment in a case of diphtheritic nephritis. Fat-grain cells and leucocytes predominating. A few red blood corpuscles.

The **prognosis** of this type of nephritis is favorable. As a rule, recovery takes place in from one and a half to two weeks, and, according to Heubner, this affection leads to the chronic form less frequently than scarlatinal nephritis. For the treatment, we refer to the chapter on scarlatinal nephritis.

(c) Nephritis Due to Other Infections, Intoxications, and of Unknown Etiology

Measles is not often complicated by nephritis, but it may appear in the early stage of the disease, and its clinical symptoms and the anatomical appearances correspond on the whole to scarlatinal nephritis. It is relatively often followed by œdema and uræmia.

Nephritis may be a complication of *varicella* too. This fact was pointed out for the first time by Henoch. The nephritis appears in the second or third week after the eruption of the papules, its hæmorrhagic character is more or less pronounced Œdema, uræmic symptoms, and even a fatal issue have been described, but the prognosis of the nephritis of varicella is in general a favorable one.

True, *smallpox* is also occasionally complicated by hæmorrhagic nephritis, but the pathognomonic importance of albuminuria appearing now and then after vaccination has not been as yet fully determined (Falkenheim).

The appearance of hæmorrhagic nephritis during an epidemic of *mumps*, or as a sequel of this disease, is rare (Mettenheimer, Henoch).

Nephritis with œdema has been repeatedly observed in the course of *whooping-cough*, and the anatomical changes have been found to consist in a degeneration of the epithelium of the urinary duct.

Typhoid in any stage may lead to nephritis, and it may be said that albuminuria is a frequent complication of the abdominal typhoid of children, appearing even on the second day of the illness.

In infancy nephritis of the hæmorrhagic type has been repeatedly observed in connection with *malaria, influenza, meningitis, pneumonia, glandular fever, erysipelas, erythema nodosum* and *general sepsis.* The nephritis which follows *tonsillitis* is also of practical importance, because it may readily be overlooked, owing to its gradual and insiduous onset. The urine contains albumin in moderate quantities, and blood, and the children feel tired and are inclined to œdema. The lingering character of the trouble is often interrupted subsequently by acute exacerbation.

General eczema is also acknowledged to be one of the causes of nephritis of older children. Guaita believes that this is the cause of the sudden deaths which sometimes occur from eczema. As to the nephritis due to *therapeutic interference*—either external or internal applications of certain medicines (balsam of Peru, styrax, tar, iodine, carbolic acid)—the remarks which were made in regard to other forms of nephritis are true in regard to this form also.

As for the more recent observations upon the damaging effects of salicylate of soda upon the kidneys of adults, it appears of sufficient interest to mention that Steffen long ago pointed out the relation of albuminuria and casts to salicylic medication.

The so-called primary nephritis of unknown origin, a class which will probably become smaller and smaller, is represented by the most varied symptoms Sometimes its character is hæmorrhagic, sometimes not. The treatment follows the principles already discussed in speaking of scarlatinal nephritis, just as in the other forms of nephritis which have been described in this chapter.

Wagner's dictum in 1882 is on the whole still valid to date. "Our present knowledge of the various forms of acute Bright's disease, is not sufficiently advanced to enable us to form a positive conclusion as to the exact etiology, either from the condition of the urine, or from the ultimate persistence of symptoms."

2. *Chronic Nephritis*

The obscurity of many points in the study of chronic nephritis in adults is still more embarrassing when we attempt to examine its pathology in infancy. A few positive signs are arrayed against many negative ones. The study of its *etiology* is also beset with difficulties. We are often in the dark as to the time of onset of the disease, because the symptoms develop imperceptibly, or are, as many writers say, chronic from the beginning. Only occasionally do we obtain a characteristic picture. In other cases they are not recognizable. At any rate we can never say that the symptoms are as definite as in the more important forms of acute Bright's disease. The *pathologic anatomy*, owing to the numerous transitions, is also ill-defined. The study of the extension, and the course of the disease is attended by great difficulties, and it is almost a lucky chance if we are able to follow the destiny of the small patient beyond the age of fourteen.

From what has gone before, it is no wonder that in the regular course, opinions as to the frequency and course differ so widely. We will only mention that for instance Baginsky emphasizes the frequency of chronic nephritis in infancy, while Biedert says that the affection is very rare. The fact, too, that the characteristic aspect of orthotic albuminuria is still historically uncertain, may contribute to the general confusion.

As a result of this condition of affairs, it will be better to base our statements upon the results of Heubner's examinations, which extended over many years, than to rely upon a literature which is so rich in contradictions. We have adopted the classification of Wagner and Heubner.

(*a*) Chronic Bright's Disease (Second Stage, Large White Kidney)

This variety seems to be the least frequent in infancy. Its course and pathology resemble in general the disease as seen in adults. The *characteristics* of the disease are scanty urine, high percentage of albumin, a plentiful sediment of all sorts of cylinders, fatty cells, renal epithelium and leucocytes.

For the **pathology** we refer to the kidney shown in Plates 49–50, which comes from a thirteen-year-old boy who died of this affection. The thickened cortex is opaque yellow in color, upon a light transparent back-ground, especially between the medullary sheathes of Bertini—a symptom of the high-grade degeneration of the parenchyma.

As in most of the described cases. the etiology in this case was obscure. In some there seems to have been demonstrated a connection with previous infectious diseases.

The **prognosis** is generally unfavorable—death takes place after months or years.

Treatment will be considered in connection with the other forms of chronic nephritis.

(*b*) Contracted Kidney (Granular Atrophy)

Heubner is of the opinion that most of the contracted kidneys which have been observed in older children are of the secondary type. It appears that the condition is not so rare as was formerly supposed. Even in 1897 Heubner was able to use for his comparative study 30 cases which had come to autopsy.

Amongst the *etiological* factors already mentioned, syphilis and scarlet fever, and heredity have been emphasized. In the majority of the cases, the exact causal factor cannot be proved with certainty.

The urine is abundant, of low specific gravity, light in color, and contains a small percentage of albumin, which disappears from time to time. The sediment, which is small in amount, contains hyaline casts.

The disease leads to retarded development, and the children feel weak and faint. Baginsky observed excessive emaciation and dryness of the skin; Förster, serious nervous symptoms, tremor and psychical depression. After a fairly long time the characteristic alteration in the vascular system (hypertrophy of the ventricle and tension of the radial artery) develop. The ophthalmoscope often leads to a diagnosis by revealing the characteristic changes in the fundus.

In its *pathological anatomy* the disorder does not differ from that of adults. The contraction of the kidney may be extreme. Most cases result in death during childhood; usually after a duration of three to four years. The patients seldom attain a more advanced age. Death occurs with symptoms of uræmia, of apoplexy, or from the hæmorrhagic diathesis. Therapeutic measures are without value.

(*c*) Chronic Hæmorrhagic Nephritis

This form, according to Wagner, is characterized by the absence of œdema, and by the occurrence of acute exacerbations, during which greater or smaller quantities of blood will be secreted with the urine, which contains albumin and casts. The quantity of urine diminishes with the appearance of blood in the urine. The **prognosis** is relatively favorable. Of six cases observed by Wagner, four recovered. It seems worth mentioning that in a series of cases the causal factor was tonsillitis (see the remarks about nephritis following tonsillitis).

(*d*) Doubtful Forms

This name was chosen by Heubner because in these cases the symptomatology does not permit them to be classed with the other forms of nephritis already described, and because the termination seems to be uncertain. All the cases continued beyond the age of infancy. Of these doubtful forms, the larger number seemed to belong to the nephritis of older children.

Etiology.—Heubner found amongst 35 cases the most frequent cause to be the infectious diseases (most often scarlet fever and then diphtheria, measles, influenza, and tonsillitis).

Symptomatology.—The symptoms bear slight resemblance to a renal disease. As a rule we find general weakness, a pale skin, the child easily tired, mentally and physicially, *i.e.*, the symptoms we are acquainted with as occurring in orthotic albuminuria. These disturbances decrease as the child grows older. Headache, vomiting, inclination to diarrhœa, are seldom observed. Heubner was not able in a single case to demonstrate dropsy, retinitis, hypertrophy of the heart, or excessive tension of the vessels. The quantity and the specific gravity of the urine is nearly normal. The daily quantity of albumin secreted is seldom higher than 1 per cent., and generally remains far below this. The sediment very scanty in quantity, contains hardly anything except hyaline casts, though there are sometimes granular, epithelial and waxy casts. Fatty cells and red blood corpuscles are absent, while the leucocytes are sometimes found in groups resembling casts. The albuminuria is often of the orthotic type. The differential diagnosis from orthotic albuminuria may be very difficult, and is frequently to be established only from the results of sedimentation. Heubner gives no information as to the cause of his cases. He refers to the observation of Aufrecht, Dixon Mann, Slawyk, who saw death occur between the ages of twenty and thirty with symptoms of uræmia in certain patients who suffered with this form of nephritis.

The **prognosis,** therefore, is not favorable, although there is no reason to abandon all hope of recovery in every case. This is shown by the observation of some cases in whom a cure was effected about the age of puberty; but the individuals who recover during this period are often left with a lowered resistance which predisposes them to relapse.

Treatment.—It is a melancholy confession to make that the treatment of the above-mentioned forms of chronic nephritis is almost useless. Only in the hæmorrhagic types is Heubner inclined to ascribe any curative value to long-continued rest in bed, absolute milk diet, and diaphoretic treatment. This is not true of the other forms, on the contrary it would be a mistake to keep patients with contracted kidneys or doubtful nephritis in bed for a long time. They simply lose their appetite and the subjective symptoms increase. These children should go tranquilly to school, and they should not be prohibited from play, work and bodily exercise; they should only be guarded against cold and overexertion. During the summer vacation a temperate mountainous region is to be preferred to a sojourn at the sea-shore. The diet should be varied, only an excessive meat diet being avoided (Weigert). Alcohol should not be given. The Carlsbad cure is worth trying, and in the case of patients who are inclined to œdema, in consequence of the large white kidney, a diet free from salt is to be recommended. In regard to the organic therapeutics recommended by the French and the Italians, we, in Germany, have little experience.

(*c*) Amyloid Degeneration of the Kidney

According to Wagner's statistics, four and one half per cent. of cases of amyloid degeneration occur in children under ten years of age.

The *etiology* and *pathological anatomy* are the same as in adults, the causes being protracted suppuration in the bones, glandular tuberculosis, malaria and syphilis. Amyloid degeneration has also been observed after a relatively short time (thirty days) in the diphtheritic kidney. The urine is light in color and its quantity is increased. The greater portion of the albumin, which is formed in abundance may consist in the albumin body precipitated by acetic acid (Senator, Joachim).

The *clinical progress* depends on the original disease. Gastro-intestinal symptoms, especially diarrhœa, are frequently present.

The **prognosis** is unfavorable, death occurring after a few months from marasmus or uræmia. Improvement is only possible if the orignal disease (syphilis, suppurations) is healed.

Diagnosis is based upon the presence of the etiological factors, together with the swelling of the liver and spleen which is usually found at the same time.

3. *Suppurative Nephritis*

This affection may originate from emboli (hæmatogenous) carried in the blood stream, from inflammatory diseases of the urinary discharging tracts (ascending), or from suppurative processes in the neighborhood of the kidneys. The hæmatogenous form is characterized by symptoms of a general pyæmia.

The **diagnosis** is suggested by the presence in the urine of casts, and masses of pus which represent a serious sepsis. If the kidneys are painful and can be palpated, the diagnosis will be easier.

The **prognosis** depends upon the nature of the original disease, and treatment must be directed to the latter. A termination in contracted kidney is possible. Renal suppuration resulting from ascending inflammation is not easily recognized because the symptoms of the inflammation of the urinary tracts (cystitis, pyelitis) dominate the picture. But the participation of the kidneys may be suspected if there appear severe toxic and septic symptoms, marked general disturbance, complete loss of appetite, vomiting, profuse diarrhœa, high remittent or intermittent fever, and if palpation shows the kidneys to be enlarged and tender. The condition of the urine hardly differs from that seen in cystopyelitis (see special article). The participation of the urine in the inflammation makes the prognosis more serious, but recovery has taken place under the same therapeutic measures which are employed in pyelitis.

TUBERCULOSIS OF THE KIDNEYS

In this place we shall discuss only those cases of renal tuberculosis which are distinguished by special local symptoms. They are in a minority, because the symptoms which proceed from the kidneys are as a rule masked by those of the general tuberculosis. If we discover the existence of tuberculosis in one or more organ, and associated with it we find albuminuria and casts it may be impossible to decide whether we have to deal with a nephritis complicating tuberculosis, or with a case of tuberculosis of the kidneys.

Renal tuberculosis is characterized by *general and local symptoms*, and by changes in the quantity and quality of the urine. The children have fever, become emaciated, and palpation reveals enlargement of the kidney, or even where the examiner is particularly skilful, the presence of tumors in the renal tissues. Pain in the kidney or tenderness to pressure are present. The former is especially apt to be found where there is perinephritic involvement, which is attended generally by spastic rigidity of the psoas muscle. The only symptoms of renal tuberculosis in younger children are incontinence and dysuria. The urine often contains great quantities of albumin, casts, pus corpuscles, especially lymphocytes, and blood. If the ureter is occluded there will be no changes found in the urine. The finding of Koch's bacillus of tuberculosis in the urine renders the diagnosis positive. If the disease is restricted to one kidney, a surgical operation may effect a cure. Ureteral catheterization, and examination of the condition of the urine from the other kidney is here of the first importance to decide whether an operation is proper or not.

STONE IN THE KIDNEY

In addition to uric acid infarction, endemic conditions have a decided influence upon the formation of a renal concretion.

This is the reason why physicians for children who have seen hundreds of cases are contradicted by those who, because they have not had the experience, believe that the occurrence of renal calculi in children is very rare. In Thuringia, and in Hungary there are regions of stone formation. Heredity, especially from gouty parents, is of importance in addition to endemic conditions. The amount of calcium in the drinking water is also considered an important factor in the etiology. Boys are more disposed to this disease than girls. The phosphates take part in the formation of renal calculi, as well as do the uric acid salts and the oxalate of calcium. Cystin stones are exceedingly rare.

Symptoms.—The concretions produce an inflammation of the renal pelvis, and symptoms of pyelitis and pyelonephritis predominate. There may be serious paroxysms of pain, which are the result of the incarceration of the stones. Their symptomatology is well known from the pathology of the adult. As a rule they are followed by hæmaturia. The diagnosis is established by the discharge of calculi in the urine, by the changes in the urine, resulting from pyelitis, by the paroxysms of pain with consecutive hæmaturia, and by the results of palpation, and of examination by means of the X-ray.

The tendency to remission of the symptoms renders the condition very chronic in its course. Amongst the complications there have been noted the formation of abscess of the kidney, with subsequent escape of the pus towards the surface, and occasionally but fortunately less frequent, into the peritoneum.

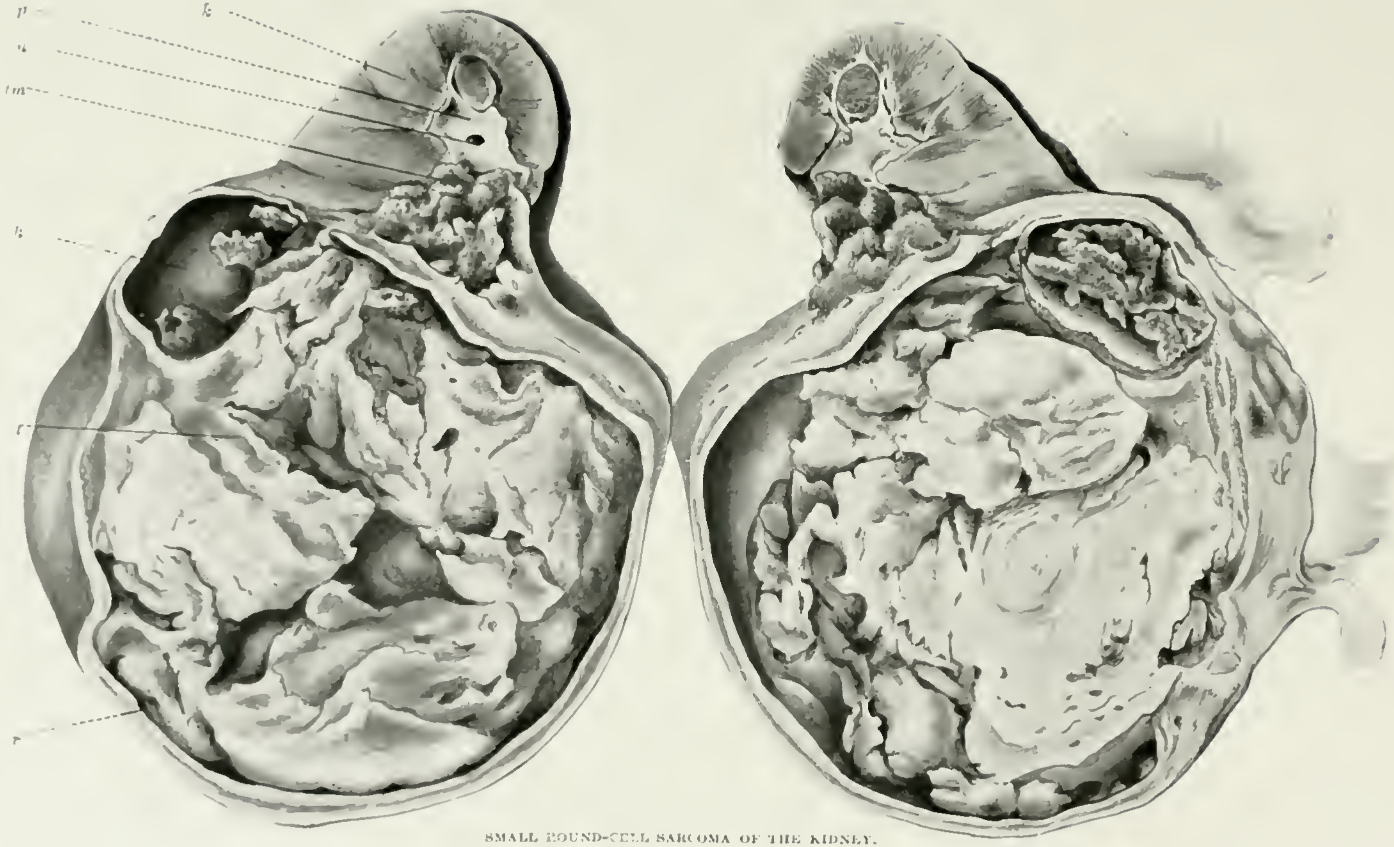
Internal *therapeutic measures* are confined to the prescription of a vegetable diet and a liberal supply of alkaline water (Vichy, Wildunger, Carlsbad water). More recently the glycerin cure recommended by Herrmann, 5–15 Gm. (1¼–4 dr.) at a dose to be taken in water, has been extolled. Urotopin fails to relieve the symptoms of cystopyelitis as long as the concretions have not been washed out or removed by some other means. Where there are symptoms of incarceration we cannot escape the use of narcotic drugs, in older children, morphine, in younger ones chloral hydrate by enema. In the present state of renal surgery we need not hesitate to recommend the operative removal of the calculi.

CYSTIC KIDNEY, WATER-BAG KIDNEY, HYDRONEPHROSIS

Cystic degeneration of the kidney is in most cases bilateral. In this condition the kidneys form a system of sacks, separated by areas of normal renal tissue.

In regard to its *genesis*, there are three opinions: Virchow believed that the cysts were retention cysts, resulting from obstruction of the urinary ducts, either by uric salts or by fœtal interstitial nephropapillitis. Two other theories assume a cyst formation upon an œdematous base, or from an arrest of development (Erich Meyer, von Dungern). The organs may be normal in size, or they may become larger than a child's head and interfere with delivery (fœtal giant kidneys, Schenkl).

PLATE 58.

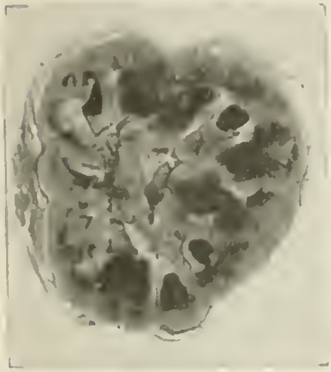


SMALL ROUND-CELL SARCOMA OF THE KIDNEY.

t, tumor, mostly necrotic; *h*, hæmorrhagic cyst; *c*, thick fibrous capsule; *p*, pelvis of kidney; *tm*, tumor extending into pelvis; *u*, opening of ureter; *k*, normal kidney tissue.

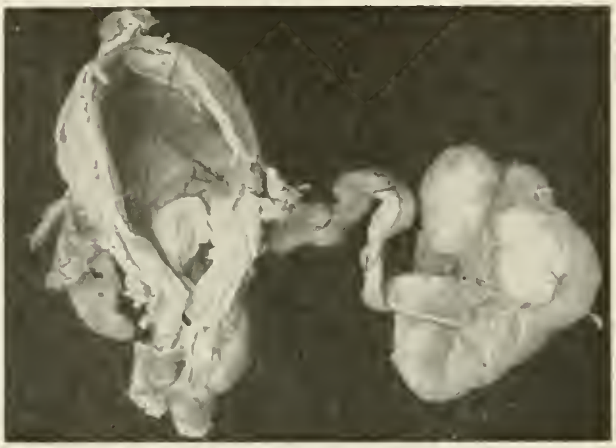
If there remains a sufficient quantity of active renal parenchyma the affected individual may live for many years; if not, death occurs sooner or later with symptoms of uræmia. The clinical course of cystic kidney may be either without any symptoms, or it may assume the aspect of a renal tumor (see Figs. 6–7). Since it is common for the affection to be bilateral, it is important to investigate the functional ability of the other kidney before attempting the removal of the cyst by operation.

Fig. 6.



Cystic kidney. System of cavities separated by parenchyma.

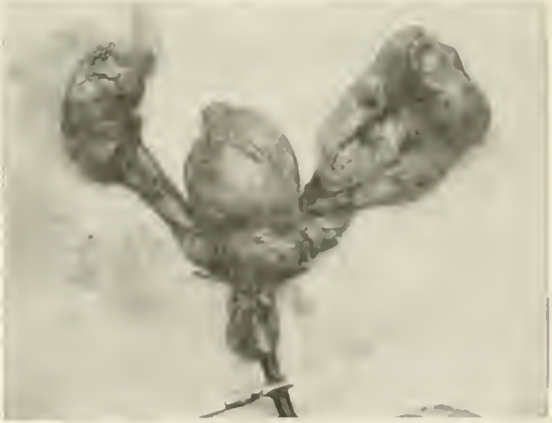
Fig. 7.



Cystic kidney with diverticulum of the bladder.

By the name of *water-bag kidney* (hydronephrosis) we understand a dilatation of the renal pelvis with consecutive atrophy of the organic parenchyma. It may be either congenital or acquired; unilateral or bilateral. The acquired cases (resulting from concretion, from inflammatory obstruction of the ureter, from compression of the ureter by tumors filling the abdomen), are probably rarer during infancy than the congenital cases. If they develop acutely, they may cause pain in the thorax and abdomen, vomiting and symptoms of urinary intoxication. The causes of the congenital type are anomalies in development of the ureter or of the urethra, which result in stricture of these tubes (atresia of the ureter, abnormal valvular formation, congenital phimosis, etc).

Fig. 8.



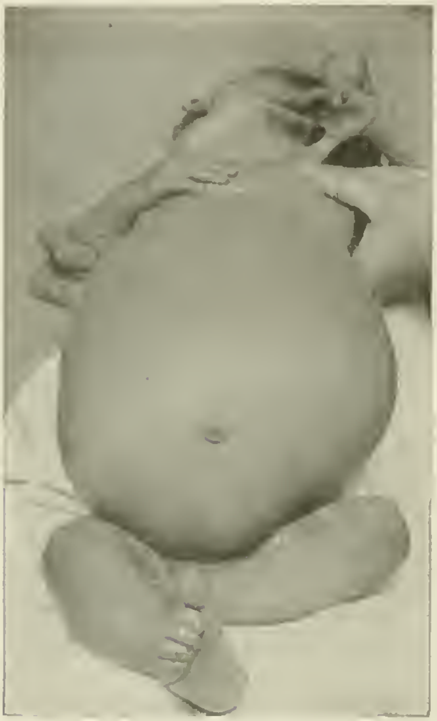
Bilateral cystic kidney, diverticulum of the bladder, dilatation of the ureters.

While water-bag kidney is not generally recognized as long as it is small, the larger ones present the appearances of a renal tumor (Fig. 9).

The diagnosis depends upon the demonstration of fluctuation, as well as upon the results of the chemical examination of the fluid which is obtained by puncture. High-grade bilateral hydronephrosis is not compatible with long life. It leads to death by uræmia.

The **treatment** of unilateral hydronephrosis is surgical. A complication which seriously endangers life, unless surgical intervention is undertaken at the right time, is the infection of the water-bag kidney, either through the blood, or through the ascending tubules, *i.e.*, the formation of a pyonephrosis.

Fig. 9.



Left sided hydronephrosis, child thirteen months old.

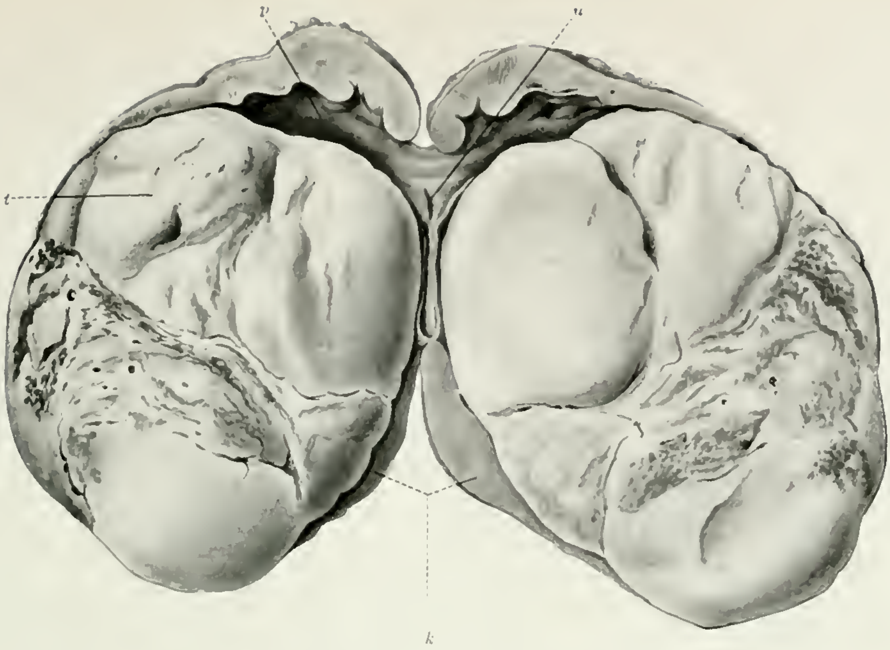
TUMORS OF THE KIDNEY

There are a number of benign tumors of the kidney (lipoma, fibroma, lipofibroma, lipomyxofibroma, etc.), which seldom attain a size greater than a walnut, and which do not produce any clinical symptoms, but which may be detected accidentally at autopsy. These same tumors, however, may originate in the renal capsule, may spread over the kidney and the suprarenal gland, and may produce the same clinical symptoms as the tumors about to be discussed in the following lines.

Amongst the tumors which have clinical importance belongs the hypernephroma (struma suprarenale, Grawitz), which originates from the suprarenal cells, as well as sarcoma and carcinoma, which Birch-Hirschfeld grouped together under the name of embryonal gland tumors. Moreover, there are found mixed tumors, adenosarcoma, myxosarcoma, lymphosarcoma, sarcoma with enclosed muscular fibres, etc. (see Plate 58). The malignant renal tumors observed in early life are supposed to be congenital in origin.

All the statistics show the remarkable fact that the greatest number of renal tumors occur in children during the first decade. Steffen compared 219 cases, and found that 34 occurred during the first year, 55 in the second, and that the sum of those occurring during the first five years was 168 out of the 219.

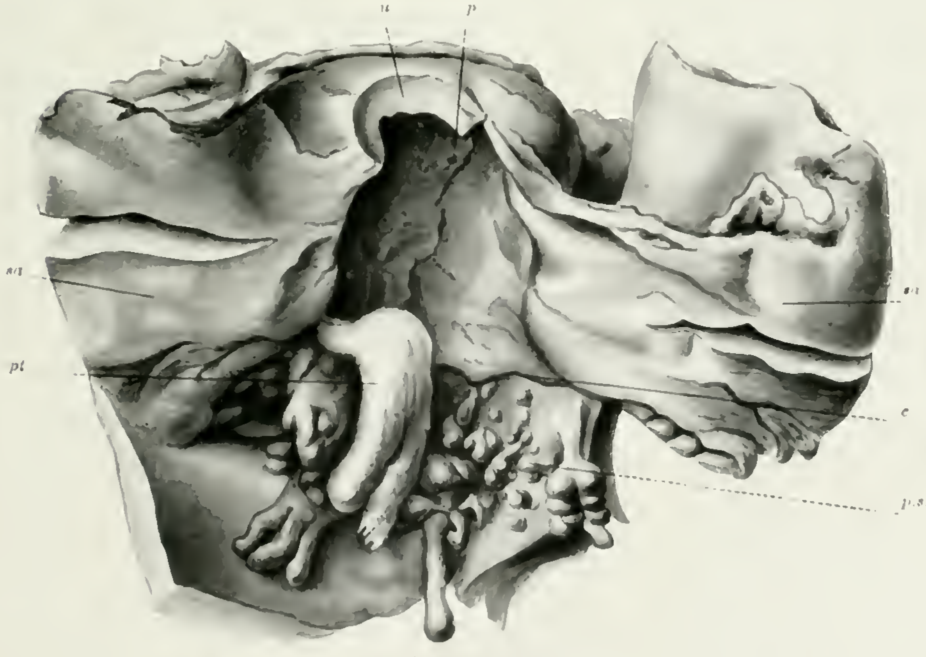
PLATE 59.



EMBRYONIC ADENOSARCOMA OF THE KIDNEY.

Removed in operation on a 6-months-old boy baby.

t, hæmorrhagic-necrotic adenosarcoma; *u*, dilated opening of ureter; *p*, dilated pelvis; *k*, kidney substance.



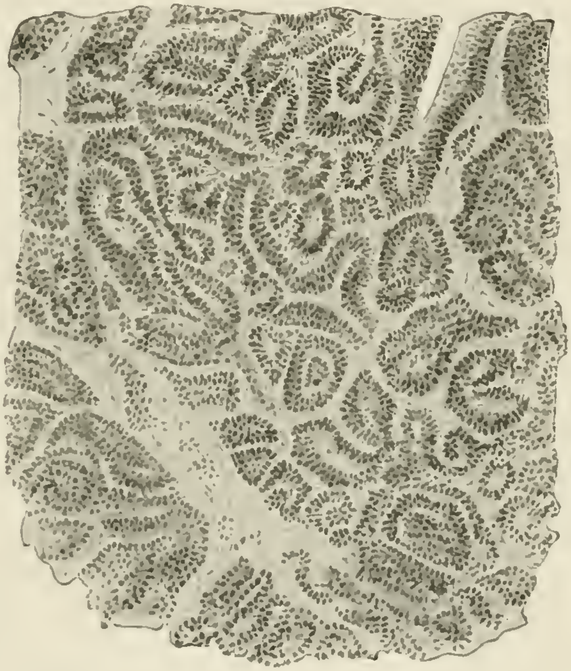
GRAPE-FORM SARCOMA.

Sarcoma botryoides of the cervix and vagina in a 2½-year-old child.

pt, primary tumor starting from the cervix; *c*, cervix; *p.s.*, polypus-like sarcomatous masses in vagina; *u*, wall of uterus; *p*, endometrium; *sa*, sarcomatous masses in lateral ligaments.

Symptomatology and Diagnosis.—It is often fairly difficult to determine whether a tumor in the region of the kidney is a renal tumor. Its relation to the ascending, transverse and descending colon is considered a matter of importance. If these parts of the intestines are distended with gas, there will be found above the renal tumor a tympanitic sound, differing from the absolutely muffled sound usually heard, but von Leube describes appearances which differ from this. Where the tumor can be distinctly outlined, and where the course has been free from fever, and moderate in its symptoms, we are inclined to exclude paranephritic abscess. A cone-shaped, blunt end speaks against an enlarged spleen, which has a wedge-shaped sharp edge. According to Israel, we may remember in differentiating between renal and suprarenal tumors, that a tumor of the kidneys is felt near the costal margin, between the ninth and eleventh rib, while the suprarenal tumor is palpable near the median line.

Fig. 10.



Section through an embryonal adenosarcoma (see Plate 52). Atypical proliferating epithelial and connective tissue strings with marked glandular arrangement.

It is frequently impossible to make a *differential diagnosis* from deeply seated tumors of the liver, or from masses of tuberculous glands, which latter must be especially borne in mind during infancy. Additional symptoms of importance are pains starting in the region of the loin and radiating towards the symphysis, and hæmaturia. The latter is said to be more common in carcinoma than in sarcoma, and may be the first symptoms of a renal nature. It is worthy of mention that Israel has described hæmaturia in connection with suprarenal tumor, which resulted from the invasion with compression and thrombosis of the vena cava and renal vein with interference with the venous drainage from the kidney. The accompanying general symptoms are emaciation, decline and gastro-intestinal disturbances The tumor still remains the most important symptom for the establishment of a diagnosis. The larger it is, the easier is it to prove its presence by palpation. In two cases Israel diagnosed by palpation, sarcomas of the size of a cherry kernel. It is exceedingly rare that any one is able to recognize tumor particles in the urine. The X-rays are of value in differentiating from calculi.

Complications result from metastatic deposits, especially in the liver and in the lungs.

The **prognosis** without surgical intervention is absolutely hopeless. Death terminates the scene either in a few months, or at least within two years. Operation which predisposes a healthy condition of the other kidney may affect a lasting cure. Out of 88 operated cases, investigated by Steffen, 18 remained permanently free from recurrence.

EPINEPHRITIS, PERINEPHRITIS AND PARANEPHRITIS

Israel understands by epinephritis an inflammation of the adipose capsule; by perinephritis an inflammation of the fibrous capsule; by paranephritis an inflammation of the loose connective and fatty tissue which surrounds the kidneys of the retrorenal fascia which envelopes the fatty capsule.

These varieties, symptomatically scarcely to be separated, form numerous transitions. In some cases the inflammation originates from a suppurative disease of the kidneys; in others, from affections of the vertebral column, psoas abscesses, pleuritis, but it may also develop primarily (Gibney, Henoch). Gibney names amongst the initial symptoms, chills, lacerating pains in the region of the loins, loss of appetite and constipation.

The **diagnosis** is established by the detection of resistance, not sharply defined, but rather diffuse, which extends from the renal region, anteriorly and towards the median line. In more advanced cases, redness, œdema of the corresponding loin, and finally abscess formation are present. The child walks in a peculiar manner, bending the body toward the affected side, dragging the lower extremity, and carrying the vertebral column stiffly and inflexibly as in the second stage of coxitis (psoas cramp). The pus may escape spontaneously to the surface, or less frequently into the pleura or the peritoneal cavity.

The **prognosis** depends upon that of the fundamental disease.

Treatment consists in incision and emptying of the pus, and in those serious recurrent cases originating from the kidney, in nephrectomy.

PATHOLOGY OF THE RENAL PELVIS AND OF THE URETER

The disposition of the renal pelvis and of the ureters may be different. There may be no renal pelvis at all, but the renal calyces may enter directly into several tubes connected with the ureter. The congenital anomalies of greatest practical importance are atresia (atresia uretero vaginalis, uretero uterina, uretero urethralis), and the formation of valves, since these conditions result in the formation of hydronephrosis.

The *prolapse* through the urethra of a blind ureteral mouth projecting into the urinary bladder like a blister, requires brief mention. More than forty such cases have been described. The prolapse may be complete or incomplete, the former being only possible in the female sex. The perfectly developed anomaly presents itself as a dark red tumor which may be the size of a hen's egg, which enlarges under pressure, is diminished by compression, and feels like a flabby bag. The latter quality differentiates it from solid tumors of the bladder. The origin of the tumor-like mass may be determined by exploration with a probe. The symptoms are similar to those of inversion and prolapse of the urinary bladder. The symptoms of incomplete prolapse, which is recognized by cystoscopic examination are like those produced by calculi (Weinlechner). In some cases it is impossible to distinguish between this condition and prolapse of the bladder. If the mouths of the ureters are visible, the former condition is present. If a kidney enlarged by the formation of a hydronephrosis can be detected by palpation, it is probable that an impervious ureter diverticulum is present.

The affection is the result of a congenital deformity of the end of the ureter.

Treatment consists in re-establishing the communication between the bladder and the closed up ureter. The danger of the condition consists in the possibility that symptoms of strangulation may appear, and in the disposition of the prolapsed membrane to inflammatory disease with extension upward.

The inflammation of the renal pelvis, pyelitis, will be discussed along with the cystitis to which the affection is closely related.

DEFECTS OF FORMATION OF THE BLADDER, OF THE URACHUS

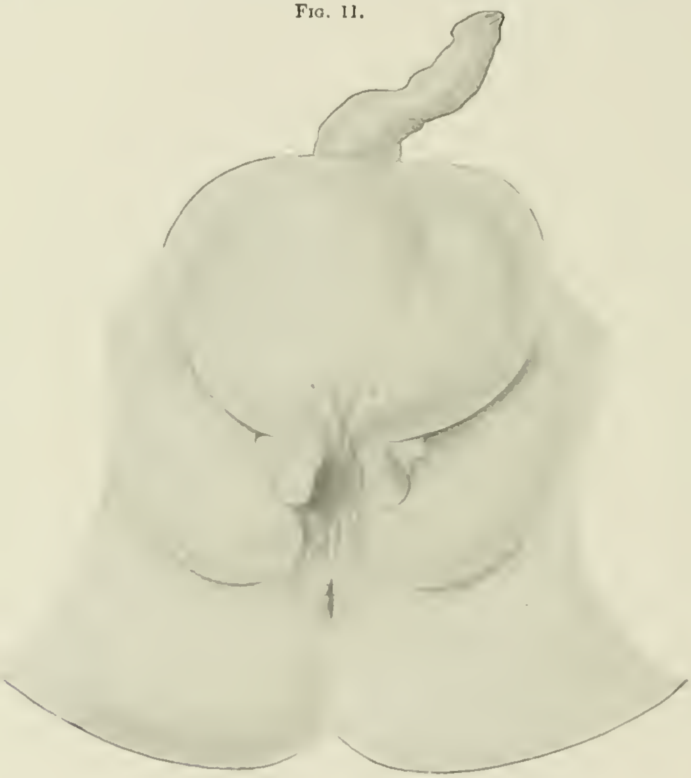
(Umbilical Fistula)

Fleury described congenital absence of the urinary bladder in which death occurred from peritonitis in consequence of catheterization, which was made necessary by incontinence of the urine. Amongst the extremely rare conditions are to be mentioned a rudimentary reduplication of the urinary bladder, with its separation into several partitions.

Ectopia of the bladder (Figs. 11, 12, and 13), which occurs predominantly in persons of the male sex, and is generally combined with deformity, is of clinical importance. Ahlfeld characterizes this affection as "a fissure in the abdomen of an otherwise well formed fœtus, which is lined with a bright red velvet-like skin (the bladder membrane), which is constantly kept moist by the urine which trickles upon it. Below the fissure in the abdomen and bladder are to be seen incompletely developed external genitals." The fissure of the abdomen and bladder may be accompanied by a complete separation of the bladder, and by an unnatural division between the two halves. To explain this anomaly, there have been brought forward theories of bursting and of incarceration.

Not only the intestines, but other organs may participate in this deformity, thus Rosenhaupt describes a fissure in the abdomen, bladder, and pubic bone with dislocation of the kidney, absence of the right umbilical artery and two separate uteri. The affection may be healed during intra-uterine life, when it may be recognized by the presence of peculiar cicatricial bands (Küster, Sonnenburg). It is of extreme danger, because of the great liability to infection of the membrane, and it should be treated by an early plastic operation. Berger proved statistically that the average duration of life is much shortened by this anomaly. Out of seventy one children, four of whom were still-born, thirty four died when less than ten years old.

Fig. 11.

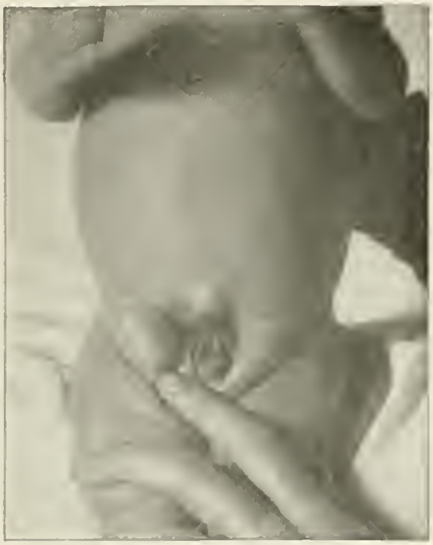


Fissure of abdomen and bladder.

Patel described a *cystic dilatation* of the portion of the urachus which communicates with the bladder, in which the bladder was pressed upon by the urachus like a tumor with resulting retention of urine.

Fistulæ of the urachus are in part congenital and in part acquired during intra-uterine life. According to Ledderhose, the first results from disturbances in the transformation of allantois into bladder and urachus. Another theory ascribes it to intra-uterine retention of the urine as a result of urethral strictures. Small umbilical fistulæ of the urachus are not so very rare. An umbilicus which remains moist for a conspicuously long time with surrounding excoriation, points to this anomaly, in the presence of which it may be possible to demonstrate a fine membrane lined with true epithelium, from which urine exudes. There often coexists deformity of the urethra (valvular blocking, strictures), and of the genitals. In undertaking to treat the condition we must consider the possibility that urethral strictures may require simultaneous removal, because after a plastic closure of the fistula of the urachus, death may result from retention and from peritonitis. Even the use of a strong caustic in the fistula may endanger life in the same manner.

Fig. 12.



Ectopia of the bladder.

INVERSION AND PROLAPSE OF THE BLADDER

Prolapse of the bladder presents itself as a dark red swelling protruding between the labia, over which the urine trickles. Predisposition to this anomaly is caused by the spindle-like shape of the child's bladder, and by the funnel-shaped passage into the urethra, in contrast to adults in whom the urethra begins suddenly as a simple round opening. The immediate **causes** are constipation, diarrhœa, and long-continued cystitis. Frua observed prolapse of the bladder in a baby six months old in the course of an attack of dysentery.

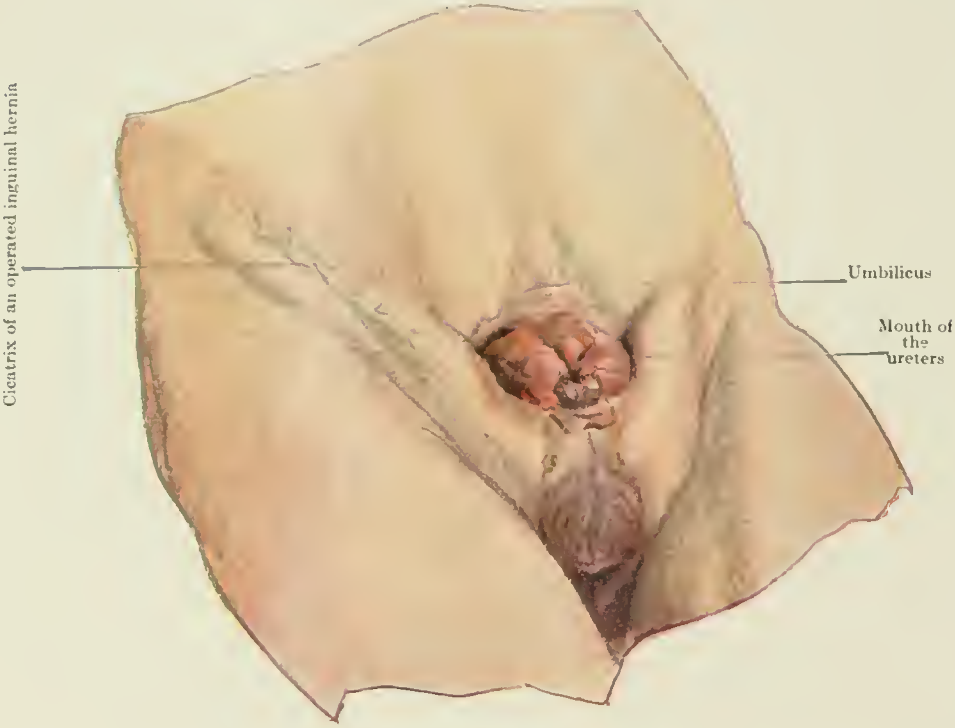
The **diagnosis** is determined by the form and consistence of the tumor, and by the possibility of replacing it, during which a greater quantity of urine is discharged (Weinlechner). After reposition we must try to preserve the position of the bladder by bandages. If we are unsuccessful in this an attempt must be made to narrow the urethra by operation.

BACTERIURIA, CYSTITIS AND PYELITIS

To Escherich is due the great merit of having pointed out in 1894 the frequent occurrence of cystitis in children, especially in girls. The bacterium coli is a frequent cause of vesicle catarrh in adults, but still more so in childhood. Escherich observed in his Clinic, amongst 60 cases that the bacterium coli alone or in mixed infection was present

58 times, and the confirmation of his observation by nearly all other writers (Finkelstein, Trumpp and others) has led to the establishment and definite recognition of the conception of colicystitis. Next to the bacterium coli, and far less frequently, there are seen as exciting causes of cystitis in childhood, streptococci, staphylococci, gonococci, proteus, bacterium lactis aërogenes, bacillus mesentericus, bacillus pyocyaneus and the bacilli of diphtheria and tuberculosis.

Fig. 13.



Ectopia of the bladder. Epispadias, cryptorchism, dislocation of the umbilicus, absence of the symphysis.

Different writers have separated the bacteriuria more or less sharply from cystitis, *i.e.*, the inflammatory reaction of the bladder membrane upon the intruded bacteria.

Krogius defines the *bacteriuria* as characterized on the one hand by the appearance of a very large number of bacteria in the freshly discharged urine, and on the other by the absence of marked inflammatory symptoms in the mucous membrane of the urinary tracts. He does not include under this affection cases in which the presence of bacteria in the urine is simply a secondary symptom of an infectious nephritis or of a general infectious disease (we know, for instance, that the corresponding bacteria are discharged in the urine in the general infection of typhoid, staphylococci, streptococci and sepsis pyocyaneus). In bacteriuria, the urine looks like a bouillon culture of bacteria. Its smell is vapid, its reaction acid, and the sediment contains nothing but bacteria. There is seldom any increase in the cellular, round cells and bladder epithelium. The only symptoms, therefore, are the peculiarities of the urine just described, and in a minority of cases, increased micturition or urinary continence. According to Mellin, who observed ten cases, this affection is frequent. The only reason why it is so rarely diagnosed is that no direct disorders result from it, and the diagnosis is arrived at only by an examination of the urine. The younger the child the greater the predisposition towards it. A great many of the cases are found in infancy. The exciting cause is generally the bacterium coli (8 out of 10 of Mellin's cases, the other two being due to the staphylococcus albus).

Other writers take a wider view of the condition, and include under the term bacteriuria cases which show general symptoms: fever, headache, pallor, vomiting and diarrhœa. Escherich is probably correct in emphasizing, on the contrary, that these symptoms suggest an inflammatory reaction of the discharging urinary tracts, and that it would therefore be better, under these conditions, to speak of them as cystitis or cystopyelitis. The pathogenesis and therapeutics of bacteriuria are exactly the same as in cystitis.

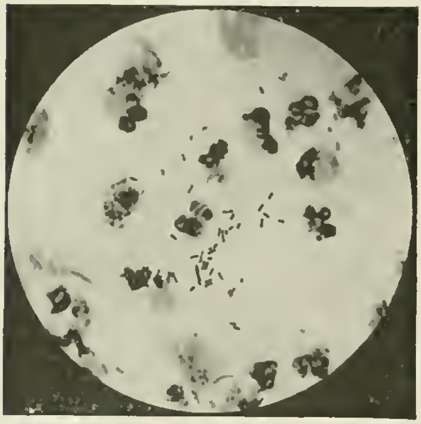
Symptomatology and Diagnosis.—Clinically we are able to distinguish two forms of cystitis in infancy. The first is attended by general symptoms, restlessness, fever, pallor, debility, anorexia; but not a single symptom pointing to a disease of the urinary tract. On the other hand the second form presents in addition to more or less general symptoms, indications arising from the urinary tract; increased micturition, difficulty in passing the urine, colic in the abdominal region, tenderness of the bladder to pressure, and inflammatory reaction in the neighborhood of the meatus. While we have to deal with obscure febrile conditions, a uranalysis is absolutely necessary where general symptoms are prominent, and local symptoms are absent, which will be the case in a large proportion of cases, especially during infancy.

In the case of infection resulting from the coli bacilli the freshly discharged urine is turbid, contains acid and albumin, but the quantity of the albumin never goes above 0.15 per cent. The turbidity is due to the presence of pus corpuscles and bacteria, which are frequently pure cultures of the bacterium coli (Fig. 14).

The proportion of the bacteria to the number of cellular elements is varied; the former are often present in far greater numbers, partly in pairs, partly grouped in small piles. Sometimes they are short like cocci, sometimes arranged in longer threads. They are readily seen in the hanging drop and are still more perceptible when Löffler's solution is added to the fresh preparation (Escherich). In order to facilitate their identification, and to prove a mixed infection, Gram's method of double staining is to be recommended, since by this process the coli bacilli are deprived of color. The bacteria will grow upon all the usual media, but there may be some slight deviations as to the growth, mobility or fermentation from the normal condition of the coli bacillus in the case of sugar solutions.

In addition to the strictly bacteriological demonstration, an acid reaction of the urine is also in favor of the presence of colicystitis. This acid reaction is constantly wanting in the case of septic cystitis due to staphylococci and streptococci, but it is present in the tuberculous form, which is extraordinarily rare as a primary affection, but which does accompany tuberculosis of the kidneys, of the genitals, or a general tuberculosis, and which can be easily recognized by bacteriological examination. This affection is said to be distinguished by severe pain, cystite douloureuse (Preindlsberger).

Fig 14.



Urine sediment from colicystitis. Smear stained with methylene blue, magnified 600 diameters.

We will briefly mention here the experiences of Göppert, who had the opportunity of observing a great number of cases of cystitis in children in Silesia. He permitted me to use a portion of his clinical experience for the present work, for which I wish here to express my gratitude. Göppert describes cases of cystitis which appeared suddenly with violent general symptoms, high fever and rapid breathing during the second half of the first year in suckling babes. Conspicuous symptoms in these cases were a characteristic pallor, appearing even on the second or third day, and loud screaming when the child was held upright. Later in the progress of the disease these infants show extreme paleness of the face, large eyes, and restless, hoarse screaming, which are symptoms recalling the aspect of severe intestinal catarrh in its later state. In older girls, Göppert saw cystitis present itself in two different forms. In one of these the child was very feverish, almost like typhoid, and there was no indication of bladder disorder. Those who suffered from the other form were brought to Göppert because of anæmia, pallor, and evening fever, *i.e.*, with the symptomatology we are wont to see in cases of tuberculosis. Vesical disturbance was present in only a small percentage of the cases.

Those forms of cystitis not caused by the coli bacillus, but by those bacilli mentioned above, differ in symptomatology very little from the colicystitis, only that their course is more malign, and that blood is more often found mixed with the urine. The pyocyaneus-cystitis which may be a complication of general pyocyaneus infection is characterized by hæmorrhages in the skin, and we can differentiate it only by bacteriological examination of the urine.

Pathologically, the acute bladder catarrh is characterized by redness, infiltration and swelling of the membrane, which is covered with mucus and with numerous cells, bladder epithelium, and leucocytes. The chronic stage shows thickening and greater puffing out of the membrane; if the process is particularly intense we see loss of substance and hæmorrhage. Except in the cystitis due to the diphtheria bacillus, we seldom see extensive fibrinous deposits.

Pathogenesis, Character, Etiology.—There has been much discussion as to the mode of entrance of the bacteria into the bladder, and as to the circumstances under which they produce inflammation of the mucous membrane. There are three possible modes of entrance; they may enter by way of the urethra; they may reach the urinary bladder through the circulation of the kidneys, by way of the urine, or finally they may invade the urinary tract, and settle upon its membrane by penetrating its walls from the adjoining viscera. The fact, already mentioned, that colicystitis chiefly affects the female sex, and that its presence is rare in male children, argues in favor of the first-named mode of infection. The bacterium coli is almost always to be found under normal conditions upon the surface of the vulva and vagina, and its migration through the short female urethra is probably fairly frequent, especially when the mouth of the urethra is open, as is readily possible in girls in certain positions. The penile urethra, however, from its very anatomical condition, would probably exclude this mode of infection.

Posner and Lewin proved experimentally the possibility of a haematogenous coli-infection of the urinary tracts. In man this method of infection must be taken absolutely for granted, but still its occurrence may be considered rare, since a frequent invasion of the blood stream by the coli bacilli is improbable, because in colicystitis the examination of the blood was almost always negative (Wunschheim, Escherich).

The third possible manner of infection, the wandering of the coli bacilli from the intestinal tract, has also been experimentally proven (Wreden). The intestinal epithelium must have been destroyed either by disease or by artificial means. Escherich is strongly inclined to acknowledge the possibility of this mode of infection in some of the cystites, particularly those in which the bladder disease follows inflammatory intestinal diseases, especially inflammatory processes in the lowest part of the intestinal canal:—"Colitis contagiosa." According to Escherich, an infection of the urinary tract in this manner is possible not only by the bacterium coli, but also by streptococci. This mode of origin, according to Escherich, is probably the rule in the extremely rare cystitis of boys.

The second question which must be answered is whether the coli bacilli or other bacteria after having gained an entrance will always, regardless of the circumstances, produce an inflammation of the mucous membrane. The pure cases of bacteriuria, prove that this is not so. We may maintain with absolute certainty, even without referring to this fact, that the bladder membrane does not always react with inflammation to entering germs. The fact that cystitis is seldom primary, but far oftener follows other diseases, especially those of the intestines, supports the opinion that in order to bring about an inflammation there must be some disposing condition present in the bladder. Guion believes that the chief predisposing factor is retention of the urine. Retention of the urine may be due, not only to local obstruction, such as strictures, diverticulæ, etc., but it may also result from conditions impairing the general health of the children, because in these cases the urinary discharge, otherwise well regulated, may be interfered with.

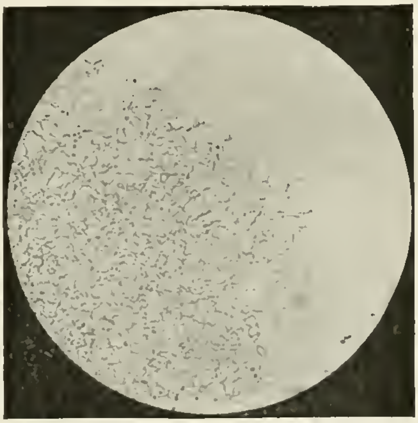
Additional factors disposing to cystitis, are cold (sitting on ice), diminished diuresis, hyperæmia, irritation and lesions of the mucous membrane by foreign bodies and by calculi, irritations by toxins, etc. (Escherich).

Our knowledge of the character of cystitis, due to the bacterium coli and the proteus has been advanced by the important discovery of Pfaundler, who demonstrated that a bouillon culture of bacilli grown on urine and mixed with the blood serum of the same patient, will produce, even if considerably diluted, the agglutination which has been proved by Gruber's experiments to occur in coli infection.

The technique employed by Pfaundler was in every essential the same as that recommended by Widal. The blood was taken by venesection and added to the serum in the usual manner. Pfaundler took the microbes he wished to examine from a pure culture (agar-agar), twenty-four hours old and deposited three drops in a bouillon tube. This emulsion he mixed with the serum in proportions of 10 to 1, 30 to 1, 50 to 1, and 100 to 1. From each of the four mixtures, and from one serum-free emulsion, as a control proof, he took one small drop and brought them together on one common sterile cover-glass, which was inverted in the usual manner over a concave glass slide. The observation was effected with a strong dry lens. Where reaction is positive after twenty-four hours, the following apperances develop: "The small rods grow out into delicate extremely long threads, which appear claw-like and interwoven and form lumpy groups under slight magnification. The groups are either isolated, or else are connected by extremely delicate filaments. Between the single filaments the liquid is perfectly free from form elements. The threads and filaments do not present the least indication of mobility. Under high powers the threads appear partly articulated, granular, and sometimes thickened into clubs. The threads are greatest in length, the filaments are densest in the reaction where the serum dilution is the least (Fig. 15).

Considering the morphological conduct described above, Pfaundler designates the reaction as the *thread reaction.* It can therefore only be called positive when it develops in a dilution of at least 30 to 1. To produce this reaction, the conditions are: The employment of a serum and of microbes from the same patient, and the presence of fever during the presence of infection as an indication of the general disturbances, but the reaction fails not only in light cases of brief duration but in serious cases which end in death. It has been established in a series of cases by means of this reaction, discovered by Pfaundler, that colon bacilli, or the proteus in the urine are not insignificant parasites, but are of pathogenic import in man, since they produce through their toxins a specific reaction in the body.

Fig. 15.



Thread reaction. Emulsion of a pure culture of the coli bacillus from the urine of a febrile child, suffering with serious colicystitis, mixed with the blood serum of the same child in the proportion of 200 to 1. Observation of the hanging drop after twenty-four hours. Magnified 150 diameters.

Course and Complications:—*Pyelitis and Pyelonephritis.*—The course varies according to the etiological factor and according to the participation in the inflammation of the renal pelvis or the kidney, for instance, if extraneous bodies or calculi are the casual factors of the cystitis, the affection will last for a time, and a cure will be effected only after the removal of the cause. Those varieties due to the bacterium coli and to the proteus, and the lactis aërogenes will be more quickly relieved than those which result from streptococci and staphylococci, whose course is more malignant. The cystites due to diphtheria, and to the bacilli of tuberculosis, belong to a more serious class, and often terminate in death.

The gradual extension of the infection through the ureter into the renal pelvis and further into the kidneys renders the course of the disorder prolonged and chronic. Symptoms indicating a secondary pyelitis resulting from cystitis, are:—sensitiveness to pressure over the renal region, radiating colicky pains in the neighborhood of the kidneys, elevation of tempertaure, which is intermittent, and "of which the type reminds us of malarial fever," with a feeling of perfect well-being during the intervals. The spread of the inflammation to the membrane of the renal pelvis is, according to Rosenfeld, recorded in the urine by an increase in the percentage of albumin, by the appearance of distorted white blood corpuscles, crenated red blood corpuscles, and the small cubical epithelium of the upper urinary tracts. The primary pyelitis which is observed especially in older girls, may present clinically the same appearances as the secondary, but sometimes not a single symptom indicates the seat of the disease, except the irritability of the child, occasional febrile reaction, an extreme pallor, and a lack of appetite, which look more like the symptoms of a general disease than of a local infection. The diagnosis can only be established by an examination of the urine, which is supposed to have, according to Graf, a smaller percentage of bacteria, a greater percentage of albumin, and a more decided acid reaction, than cystopyelitis.

If the kidney becomes involved in the inflammation, it will be shown, as in the case of an infant whose cystitis followed upon an intestinal disease, by exacerbation of the gastro-intestinal symptoms, by the appearance of vomiting and diarrhœa, which do not depend upon the character of the nourishment, and which alone point to the nature of the disturbance. The percentage of albumin in the diminished urine is increased, and casts and renal elements appear. Convulsions, opisthotonos, and coma may be added to the picture and the patient may die with high remittent or intermittent fever, with symptoms due partly to the urinary intoxication, and partly to the septic infection.

If we bear in mind what was said of the symptomatology of the nephritis complicating the gastro-intestinal diseases of infancy, it is apparent that where a cystitis has come on rapidly in the course of an intestinal disease, it is hardly possible to establish the differential diagnosis between cystitis associated with a simulanteous hæmatogenous nephritis, due to the fundamental disease, and a pyelonephritis. Only the postmortem will certainly clear up our doubts in many of these cases.

Prognosis.—With proper treatment the prognosis of cystitis, except in the forms resulting from diphtheria and the tubercle bacillus may be considered in general to be favorable. Without medical treatment some of the cases, especially in infancy, terminate in death with septic symptoms. Even involvement of the renal pelvis does not greatly increase the seriousness of the prognosis, but where the infection spreads to the kidney the danger of the outlook is greatly increased.

Prophylaxis and Therapeutics.—Cleanliness of the genitals is of the utmost importance in prophylaxis. Those who have the nursing of children should be directed to bathe the anal and the genital regions only from the front towards the back, since otherwise the bacteria about the anus may readily be introduced into the urethra. Therapeutic measures are directed to the cause. In the presence of conditions which excite or maintain a cystitis, of anomalies of the urinary tract, of extraneous bodies or calculi, we must, in the first place, treat or remove these causes. Only after they have been gotten rid of can medicinal and dietetic therapeutics be effective.

The sovereign remedy for cystitis and pyelitis is urotropin (hexamethylenetetramin) introduced by Nicolaier, which, when taken by the mouth counteracts the influence of the exciters of the infection by splitting off formaldehyde during its excretion through the kidneys. For infants 10 c.c. (2½ dr.) of a solution of 13 Gm. (3⅓ dr.), of urotropin in 100 Gm. (3½ oz.), of water should be given three times daily in milk. To older children larger doses up to 1.5 Gm. (25 grains) per day can be given. Instead of urotropin, helmithol has been recommended, but it possesses no superiority, and children dislike it because of its disagreeable taste.

Another remedy which is fairly popular, and quite effective, is salol. The doses should not be too small if we are to obtain good results. For infants we should use 0.1 to 0.3 Gm. (1½–4½ grains); for older children 0.5 Gm. (7 grains) four times a day. Recovery will be hastened by simultaneous irrigation of the bladder, which is to be effected in babies by introducing a small metal catheter, which is fastened by a short piece of rubber tubing to a syringe holding 10 c.c. At first a 3 per cent. solution of boracic acid is used, 3 to 5 injections are given, after each of which the boracic solution is allowed to escape again and then in the same way three injections of a solution of nitrate of silver (1 to 2000 to 1 to 1000) are introduced, and finally the remnants of the solution of silver are precipitated by the introduction of 3 to 5 syringefuls of a 1 per cent. solution of common salt. Violent pain may necessitate the symptomatic use of poultices and of narcotics; nausea and vomiting are effectively combated by cracked ice.

The nourishment of the infant during this affection is indicated by the condition of the child. Food rich in salts is to be avoided, and liquids are to be supplied in abundance. Older children should be given a diet free from spices and consisting of milk and vegetable dishes. Drinks which may be given are: whey, fruit lemonade, almond milk, or alkaline waters. The children must be kept in bed until the acute symptoms, fever and pains, have disappeared. They need careful nursing, and must be guarded against any bodily exertion or catching cold.

PERICYSTITIS

Gallasch described suppurative inflammation in the tissues around the bladder in infancy as a primary process. Besides a general feverish condition, the symptoms consisted of frequent micturition, pain above the symphysis, and a bogginess in this region, not affected by changes of position. The differential diagnosis from a localized peritonial exudation may be very difficult. In the case described by Gallasch, recovery occurred after rupture of the abscess into the rectum.

STONES IN THE BLADDER AND IN THE URETHRA

What was said in regard to the influence of endemic conditions upon the appearance of stones in the kidney in infancy, is also true in regard to concretions in the bladder and in the urethra. There exists along the lower part of the Danube an area of stone formation. Bókay has been able to collect in this region alone the records of more than 1621 cases of vesical calculi. In Russia, France, and England, too, the formation of concretions in the bladder appears to be much more frequent than in Germany, where the affection is rare. Bókay's statistics show that the majority of cases occur between the second and the seventh year. The youngest child in whom Bókay observed stone formation was in its second month. Only four per cent. were girls. We learn from the tables of Englisch in regard to stones incarcerated in the urethra that more than one third occurred in children, and that most of the cases were observed in the second, and between the eleventh and fifteenth year.

Etiology.—Bókay regards interference with the urinary discharge as a factor disposing to the formation of calculi in the bladder; hence the predisposition caused by phimosis and the relative frequency of the condition in boys. The immediate cause of the stone formation may be either a concretion descending from the kidney, or a foreign body introduced for the purpose of masturbation. If there results an inflammatory reaction of the vesical mucous membrane and with it decomposition of the urine, the result will be the formation of a calculus by the precipitation of insoluble phosphates.

Englisch explains the great frequency of urethral calculi in the second year by the congenital narrowness of the urethra. In the eleventh and fifteenth years by the abundance of blood vessels and the swelling which is associated with it. Uric acid and its salts, phosphates and oxalates, take part in the composition of calculi.

Symptoms.—The symptoms are, at first, increased micturition, radiating pains in the glands and vesical tenesmus. Incontinence of the urine may be also an early symptom. Gradually there are added the symptoms of vesical catarrh with admixture of blood in the urine. The more severe the pains and the cystitis, the greater the systemic disturbance. Among the serious complications are to be mentioned deep ulcerations of the vesical membrane, with consecutive pericystitis or pelvic abscess. Prolapse of the rectum is frequently seen.

The **diagnosis** is made from the above symptoms, the most important of which is the variable difficulties in urination. At one time there will be no trouble and at another urination is painful and difficult. A correct diagnosis can only be made after a rectal examination, exploration with a sound and a cystoscopic examination.

The **prognosis** is influenced by the extension of the inflammation due to the presence of the stones in the urinary tract, and by the amount of systemic disturbance.

Treatment is surgical. Excellent results have been obtained both by lithotripsy and by suprapubic cystotomy. After the removal of the stone the inflammation of the urinary tract must be treated according to the usual methods.

TUMORS OF THE BLADDER

The bladder may be the seat of either primary or secondary tumor formation. Steffen's statistics show that the more malignant tumors of the bladder are rare in comparison to the other vesical diseases. Steffen analyzed 32 cases, the greater number of which occurred in children between one and five years of age, and the youngest of which was a child of eleven months. The majority of the tumors started from the mucosa between the openings of the urethra. Sarcomata prevailed. The tumors in the beginning without any symptoms, are characterized as they advance in development by disturbances in the passage of the urine, and by pains in the bladder. The urine contains pus and blood, rarely particles of the tumor. Hæmaturia appearing after the correct introduction of the catheter into the urinary bladder is regarded as an important symptom. Sometimes it may be possible to palpate a tumor through the rectum. If it appears at the vulva, the diagnosis may be established with certainty. The complications are hydronephrosis, pyelonephritis and suppurative peritonitis. The prognosis, in spite of operation is unfavorable.

ANOMALIES IN THE URINARY DISCHARGE

These may be either purely functional, or they may be the result of organic disease. In this place we shall speak only of the latter form.

Anuria, which has been already mentioned as a symptom of nephritis, also occurs congenitally. For instance, in deformities of the discharging urinary tracts, and in cystic kidneys, Bömann describes such a case which lasted nineteen days.

Pollakiuria is physiological near the end of the first and the beginning of the second year, and it is only at the end of this time that the discharge of urine begins to be dependent upon the will. Pollakiuria sometimes appears in older children temporarily after the acute infectious diseases. It may be a symptom in diabetes mellitus, diabetes insipidus, contracted kidney, and in irritating conditions of the urinary passage.

Retention of urine, the collection of urine in the bladder until it is distended to its utmost capacity, may occur in the first place in all those conditions in which micturition is accompanied with pain, and it may result from deformities and anomalies of structure, congenital strictures, duplications of the mucous membrane of the urinary tract, calculi, inflammations of the kidney, bladder, prostate, urethra, or prepuce, in pericystitis, congenital hypertrophy of the prostate, foreign bodies and new formations in the urinary organs, prolapse of the vesical mucous membrane, paresis of the bladder from stupor in the infectious diseases, in organic diseases of the brain and spinal cord, and further, by mechanical and reflex disturbances or new growths in neighboring organs (for example, Bartenstein described a case of retention of urine in a female infant ten months old, which resulted from a periproctitic abscess). Retention of urine may also be imitated in babies, as in children suffering from serious gastro-intestinal diseases who take but little nourishment and suffer with numerous watery evacuations (Bartenstein). Hagenbeck-Burkhardt mention the inability of children suffering with tetanus to discharge the urine, owing to spasm of the sphincter vesicæ. This subsides along with the other symptoms of tetanus.

Incontinence of urine, the involuntary discharge of the urine, may be the result of affections of the bladder (calculi, cystitis, bacteriuria), and of deformities of the genital apparatus. As causes of incontinence of urine, excoriations and irritations near the urethra (oxyuris) may be mentioned. Ray goes so far as to maintain that the "enuresis" is in most cases a symptom of co-existing cystitis, or at any rate the result of irritating conditions persisting after the cure of a catarrh of the bladder (phosphaturia, ammoniuria, gravel, excessive secretion of uric acid).

The **treatment** of these conditions is directed to the cause. Where there is a retention, the catheter must be used to relieve the symptoms resulting from the retention and the dangers of the systemic disturbance.

DISEASES OF THE MALE GENITALS

EPISPADIAS

Epispadias is an abnormal opening of the urethra on the dorsum of the penis. If the opening is on the glans we speak of epispadias glandis; if the dorsum presents a deep furrow, we speak of penile epispadias; and if, instead of the symphysis, we find a pit-like deepening, we speak of epispadias with exstrophy of the bladder.

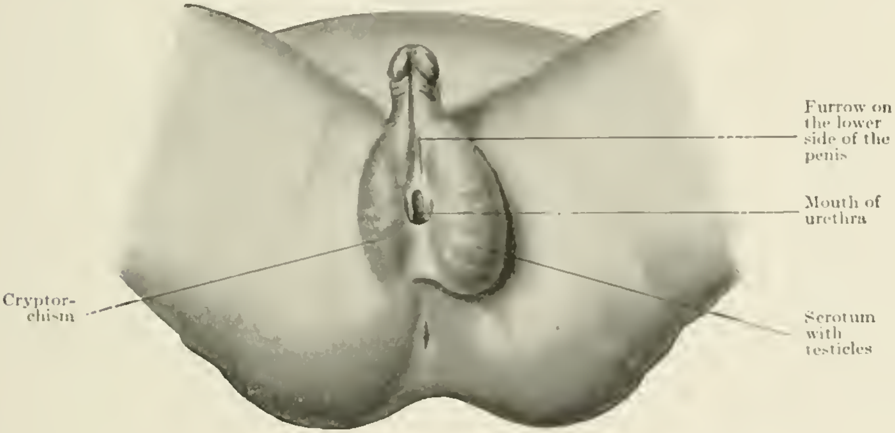
The anomaly is very rare, but it is frequent where there is simultaneous atrophy of the bladder (Fig. 12). The criterion of the anomaly is the discharge of urine from an abnormal opening upon the surface of the penis. According to one theory it is the result of arrested development. According to another, it is due to rupture of the fully developed urethra by urinary retention (see theory of bladder fissure).

The most frequent **symptom** is incontinence of urine. Among the associated disturbances are to be named eczema, a disposition to inflammatory processes of the discharging urinary tracts, and at a later age disturbances of the sexual functions. (Concerning the plastic operations see works on Surgery.)

HYPOSPADIAS

By this name is designated the anomaly in which the urethra does not open at the point of the glans, but at some other point upon the lower side of the member. If the urethra opens at the base where the frenulum normally adheres, we speak of glandular hypospadias. If the urethra opens at any other point upon the lower side of the penis in front of the scrotum, we call it penile hypospadias or peni-scrotal hypospadias.

Fig. 16.



Hypospadias.

The highest degree of this anomaly is presented by perineal hypospadias (Fig. 16). These cases may look as if the individual were feminine, because the member is rudimentary in most cases. The discovery of testicles confirms the diagnosis. If these did not descend, the differential diagnosis between male and female may be impossible.

The **etiology** of the deformity consists in a failure of the urethral end to develop, thereby forming only a furrow (the cyst formation in the median line of the raphe of the external genitals, and the presence of penile fistulæ described by Englisch are connected with incomplete union of the genital folds). Glandular hypospadias represents an arrest of development at the third to the fourth fœtal month. Penile and perineal hypospadias correspond to disturbances at an earlier stage.

The passage of urine is usually rendered difficult in some cases to such an extent that complete atresia may be immitated. Incontinence in these cases, according to Karewski, is due to the dropping of urine from the overflowing bladder.

The **diagnosis** is usually easy. In cases of seemingly complete retention of urine the differential diagnosis must be made from absolute atresia of the urethræ. The finding of even a pin-point urethral orifice will reveal the true conditions (the treatment is purely surgical).

CONGENITAL ATRESIA OF THE URETHRA

Karewski with Englisch, distinguishes in addition to the rare occurrence of complete absence of the urethra and penis, and the complete absence of the urethra with existing penis, between closure of the external urethral orifice by adhesions of the membrane; imperfect glans characterized by the fact that the glans has either no sign of a urethra at all, or only a shallow blind dimple; and more extensive obstruction of the urethra in which the entire urinary tube, or a part of it may be lacking. These disturbances cause the formation of umbilical-urachus fistulæ. The symptoms are those of retention of the highest grade, which in children born alive, is explained by the absence of discharge of urine which will soon be discovered. The life-saving treatment of this condition is surgical, and must be applied immediately after delivery.

STRICTURES AND DIVERTICULA OF THE URETHRA

Among other causes of retention of urine congenital narrowing of the external orifice is of importance. Cylindrical stricture was found in one case by Demme. Membranous duplications, producing narrowing of the canal may be present in the fossa navicularis, and in the prostatic portion. All of these affections present as a common symptom retention of urine, which may be cured by mechanical dilatation of the strictures (in Demme's case by the use of laminaria).

The formation of diverticula of the urethra is rare. We understand by this term inlets which are connected with the urethra, Bókay distinguishes between true and false diverticula. If the wall is lined with mucous membrane, he speaks of them as true diverticula; if lined with new formed tissue, as false ones. The true may be either congenital or acquired; the acquired are due to urethral calculi or to organic strictures. False diverticula appear if an abscess is formed in the neighborhood of the urethra by strictures, injuries to the inner urethra, or by artificial interference, if the abscess breaks through the wall and forms a sack; or if an abscess is formed near the urethra, but independent of it, by an external trauma and becomes a urine-containing bag. Diverticula seldom result from retention cysts of Cowper's glands.

In none of the congenital cases does the diverticulum extend further than the peni-scrotal raphe, while the acquired ones as a rule appear in the perineal region, and only exceptionally near the glans. Kaufmann names as *causes* of congenital diverticula:—disturbances in the juncture of the glands and penile urethra at a time when urine has already begun to overflow from the bladder. This will cause retention of urine, and a dilatation of the lower urethral wall where it is most yielding. In all the cases that have been observed up to this time, the lower wall was affected. The symptoms consist in the swelling or formation of a bag when the urine is discharged, and when the water is retained in it. The urine may be forced out by pressure. In some cases we find a constant dripping of urine. The treatment consists in operative removal of the diverticula.

ANOMALIES OF THE PREPUCE; PHIMOSIS AND ITS COMPLICATIONS

(Balanitis, Balanoposthitis, Paraphimosis)

The development of the prepuce and of the anterior portion of the urethra takes place in the third to the fourth month of embryonal life. During this time a fold which lies at the posterior edge of the developing glans grows over it and covers the glans completely in the fifth month.

Congenital defects of the prepuce often appear in families in which preputial deformities are hereditary. They may either appear alone, or associated with other deformities of the genital apparatus.

According to Bókay and Kaufmann, in the normal newborn child the glans is always adherent to the inner membrane of the prepuce. The space between the glans and the prepuce is filled by an S-fold layer of pavement epithelium, whose cells reach into the canoe-like pit of the urethral mouth.

The epithelial agglutination becomes loosened by movements during the growth of the first months of life, but it often takes some years before the prepuce can be completely pushed back. Bókay distinguishes three degrees in the process of loosening. In the first, when the prepuce is gently retracted the urethral orifice will be just visible in the opening. In the second the prepuce can be drawn over the middle of the glans, but is arrested at a point where they are still grown together. In the third degree the only adhesions which remain are in the retroglandular sulcus.

Theoretically there is a distinct difference between the epithelial agglutination of the prepuce with the glans, as a physiological condition, on the one hand, and the narrowing of the prepuce which prevents the glans from passing through it on the other, aside from the epithelial agglutination which would render this impossible. But in practice this difference cannot be always maintained, since the symptoms are frequently the same. Hofmokel distinguishes four causes of phimosis: (1) a prepuce congenitally too long and too narrow (hypertrophic form); (2) congenital narrowness, restricted to the external opening of the prepuce; (3) long persistence of extensive epithelial agglutination between glans and prepuce, (4) congenital and abnormal shortness of the frenulum and its location too far towards the front. In all of these forms the internal membrane of the prepuce appears shorter than the external. Karewski describes, in addition to these forms, the cicatricial phimosis appearing after birth, and the form, rare, it is true in children, which results from œdematous swelling in consequence of acute inflammation, but showing a normal prepuce.

Symptoms.—Frequent and painful urination are the symptoms of the affection. Mothers often refer the restlessness of a child to difficulty in passing the urine, and ask for an operation upon the narrow prepuce. A careful examination often shows that the connection traced by the mother does not exist, and that the restlessness of the child will disappear when, for example, the nourishment is properly regulated. Any one who has the opportunity to witness the passage of urine in babies with phimosis would observe before the action great restlessness, reddening of the face, and eventually violent screaming; and then the urine will be suddenly discharged, or a part flows into the prepuce and distends it like a balloon. In severe cases, the prepuce forms a kind of urine reservoir, out of which the retained fluid constantly drips.

The local results of the conditions are eczema at the urethral orifice, which may spread over the skin of the entire genital region; balanitis, balanoposthitis, the formation of concretions in the stagnant and thickening preputial secretion (preputial calculi).

The *balanitis* is a superficial inflammation of the glans with the production of abundant pus, mixed with the epithelium and creamy secretion of the glands. The affection is usually associated with inflammation of the inner preputial membrane, posthitis, and thus becomes a balanoposthitis. In the presence of this affection the prepuce is swollen and reddened, the membrane is ulcerated in some places and secretes an ill-smelling pus. In cases of greater intensity the swelling becomes more extensive, the exudation profuse, and gangrene may result.

It is evident that the opening of the prepuce becomes still narrower as a result of these inflammatory conditions, and that the urinary disturbances increase, a harmful vicious circle having been formed.

Further complications are, ascending cystitis, dilatation of the urinary bladder and the formation of hydronephrosis; in consequence of the forced action of intra-abdominal pressure there may appear a hydrocele, intestinal rupture and rectal prolapse. Even death by uræmia ultimately dependent upon phimosis has been described. Furthermore, a whole series of nervous disturbances, syncopes, and epileptiform convulsions, have been attributed to the presence of phimosis. The irritation of the external genitals produced by these conditions have resulted in masturbation even during infancy.

If the prepuce is forcibly retracted over the glans, the eventual result will be the disagreeable complication called *paraphimosis*, the strangulation of the penis by the narrow ring of the prepuce in the coronary sulcus. The immediate result is violent pain, interference with the circulation, the appearance of œdema, and, by the development of inflammation, the occurrence of ulceration, phlegmon, and even gangrene of the glans.

Diagnosis.—We must decide whether the case is a simple epithelial adhesion without stenosis, or a real narrowing. The history given by the mother is not to be considered of any value in diagnosis.

If there exist symptoms of inflammation of the prepuce, the urine should be examined without fail, since, as Rey has emphasized, a cystitis may have caused this inflammation which imitates a phimosis.

Treatment.—The removal of the phimosis may be effected by either bloodless, or bloody means. The bloodless method, according to Karewski, consists in freeing the epithelial agglutination by means of the flat end of a probe, and the gentle and careful retraction of the prepuce over the glans. This must be effected with careful asepsis to avoid secondary inflammation, which leads to the formation of scar tissue. Karewski advises this method only in the cases of epithelial adhesions, while for the removal of phimosis he proposes the operation of circumcision which is absolutely without danger when performed aseptically, and which is accomplished in a few minutes. (For a description of this operation see works on Surgery.)

The relief of *paraphimosis* is effected by reposition of the strangulating ring. The reposition is effected by embracing the strangulating ring with two fingers and trying to push it back, while at the same time the thumb presses the glans into the prepuce. The reposition will more readily succeed after incision of the compressing preputial ring. Cold compresses, containing alum, will soon relieve the œdema and the inflammatory irritation, and after this the operation for phimosis may be undertaken.

URETHRITIS

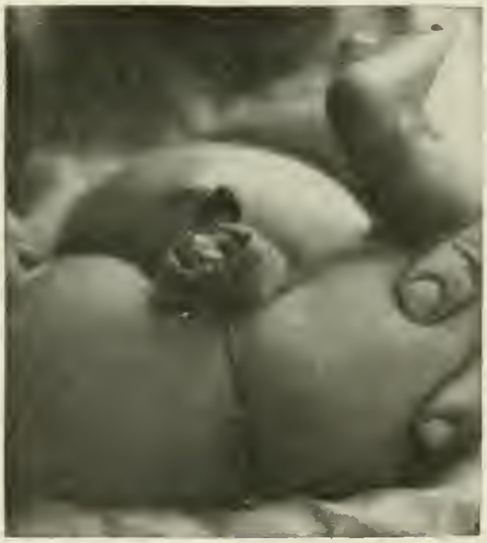
Inflammation of the urethral membrane in boys may result from extension of an inflammatory process from without. It may be caused by trauma, by the introduction of foreign bodies for the purpose of masturbation, and rarely, but still too frequently, it may be the manifestation of a gonorrhœal infection. The latter, according to Fischl, is often produced by a balanitis and is accompanied by far more violent general symptoms, as in the case of adults. It also spreads to the prostatic region, and leaves behind it in many cases a stricture.

The **symptoms** consist of pain upon micturition, in the appearance of a discharge which is sometimes chiefly mucus, sometimes purulent; of redness and swelling of the urethral orifice. The complications are, the spreading of the inflammatory process to the mucous membrane of the bladder, and those complications of gonorrhœa which will be spoken of in discussing vulvo vaginitis. The inquiry into the etiology is of importance in determining the treatment. A urethritis due to balanitis is soon relieved by treatment of the latter, by removal of the secretion, and the establishment of a free discharge by a tampon saturated with an astringent liquid. If the gonococcus is the cause of urethritis it must be opposed by the usual injections of a silver solution.

GANGRENE OF THE SCROTUM

This affection is usually the result of a deep-seated inflammatory process which has spread to the scrotum. It may originate from an erysipelas of the abdomen or thigh, from suppuration of the inguinal glands, from inflammation of the prepuce. Sometimes it will result from a urinary infiltration caused by traumatic rupture of the urethra. Less frequently its origin is enigmatical (Fig. 17).

Fig. 17.



Gangrene of the scrotum.

The local **symptoms** are, in the beginning, high fever, a hard infiltration of the bluish red colored scrotum with reactive inflammation and œdema of the neighboring parts. Gradually there appear isolated ugly colored places, which soon become confluent, and of which the tissue dissolves into a foul smelling mass. At the same time, there are exceedingly severe general symptoms. Coma and convulsions are not rare. The termination is often unfavorable; the children dying of cardiac weakness with symptoms of general septicæmia. In the few favorable cases, there was demarcation of the gangrenous tissues with shedding and healing by cicatrization.

Therapeutic measures are confined to efforts to assist the exfoliation of the gangrenous areas, and to preserve the strength of the child by a concentrated diet and by stimulants. Prolonged baths with antiseptic solutions will soothe the pain.

ANOMALIES OF POSITION OF THE TESTICLES

(Retentio testis or cryptorchism. Ectopia testis. Inversio testis)

The descent of the testicle, illustrated in Fig. 18, should be completed during the eighth fœtal month. If they are arrested **in** their progress we speak of a retentio testis, of which there are two degrees: (1) *abdominal retention,* the cryptorchism in which the testicles remain within the abdominal pelvis, and (2) the *inguinal retention* in which they are arrested in the inguinal canal. It is a remarkable fact that the completion of the descent of the testicles may be accomplished even late in childhood. Soltmann observed two cases in which the descent occurred in the ninth, and eleventh years, and Velpeau a case in which it took place in the twenty-second year. In cryptorchism the testicles can neither be felt in the scrotum, which is somewhat backward in development, nor in the course of the inguinal canal. In "retentio inguinalis," the testicles can be felt in the opening of the inguinal canal, as a small oval body, with its long axis corresponding to the fold of the groin (Soltmann).

The **differential diagnosis** must be made from a small hernia. Cryptorchism seems to be comparatively frequent in the newborn: among 102 newborn boys, Kiebert observed 30 cases of unilateral or bilateral cryptorchism. Later on the disturbance often is partially relieved, and the percentage becomes smaller (among 10,800 cases it was found 12 times).

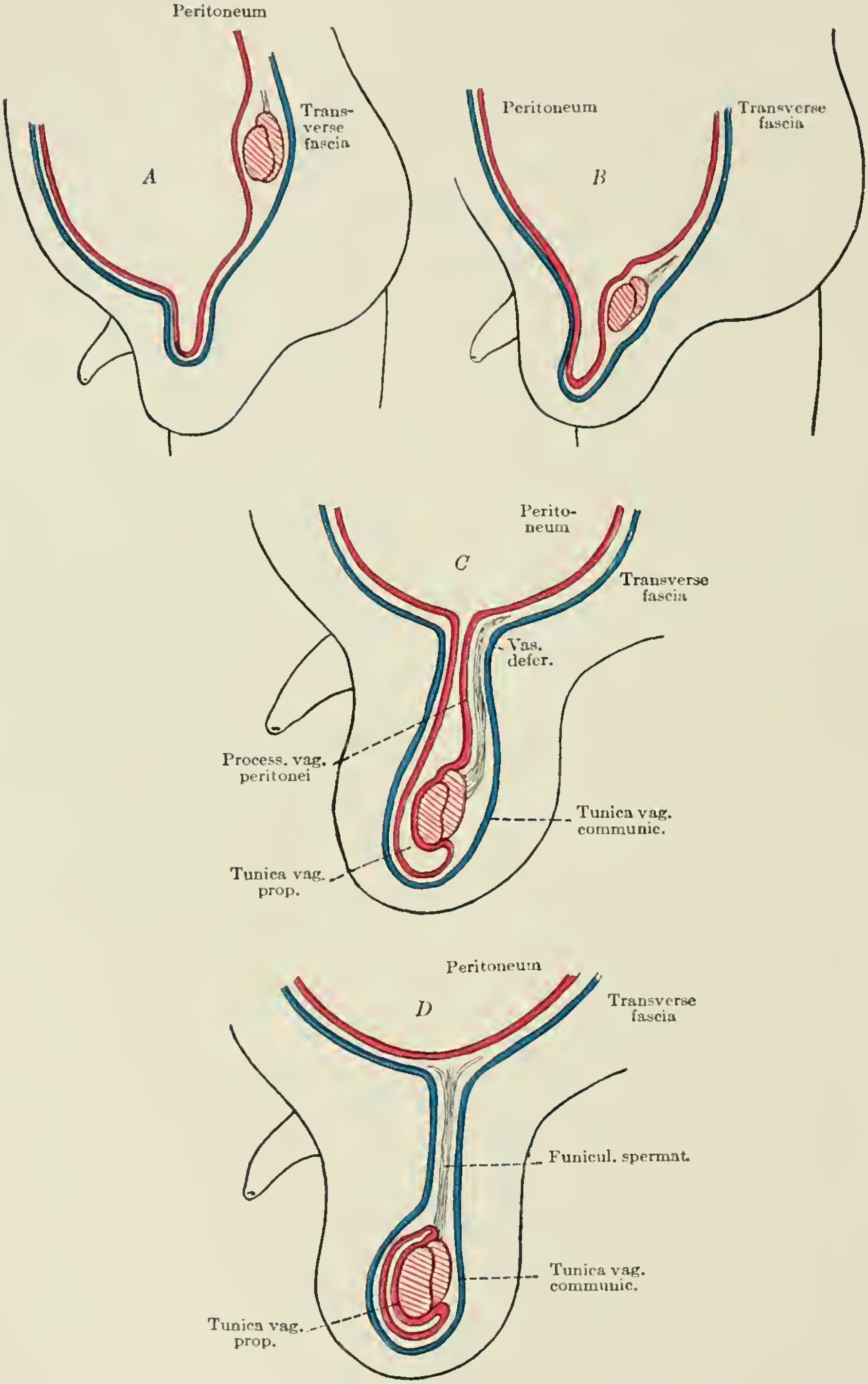
Amongst the **etiological** factors are to be mentioned: abnormalities of the gubernaculum, narrowness of the inguinal canal, intra-abdominal adhesions of the testicle (adhesions of the left testicle to the sigmoid flexure due to fœtal peritonitis, in one case described by Tandler).

Non-descent of the testicles is of clinical importance because it creates a disposition to inflammatory processes, whose course may be so violent that they remind us of the symptoms of an incarcerated hernia. Cryptorchism is often followed by the appearance of herniæ, and it also favors the development of incarceration.

Among the **complications** must be included: hydrocele, torsion of the spermatic cord, with gangrene of the scrotum. By extension to the cavity of the peritoneum, inflammation of the inguinal canal resulting from trauma may lead to peritonitis. Among the sequelæ mention must also be made of atrophy of the testicles, and a tendency to malignant degeneration of the organ. According to Soltmann, atrophy of the testicle is not the immediate result of cryptorchism, but is brought about by a chronic inflammation of the testicle, which may be due to trauma from without, or to strangulation from within. In addition to these purely local disturbances there are more general symptoms, such as syncope, hystero-epilepsy, which have been referred to this anomaly of position.

By way of **treatment**, Soltmann recommends that the descent of the testicle be encouraged by massage, and if this is successful, the fixation of the organ by means of proper bandages. Other writers prefer to operate, freeing the testicle, and anchoring it in its proper position. Sebileau advises that no interference be taken in early infancy, but that massage be used for two to five years, and that operation be postponed till near the age of puberty.

Fig. 18.



Descent of the testicles. *A.*—Position of the testicle about the 4th fœtal month. *B.*—About the 6th to 7th month. *C.*—About the 9th fœtal month (appearance of process. vaginal. peritonei). *D.*—Position of the testicle at delivery (appearance of the tunica vaginalis propria).

Of lesser clinical importance are the anomalies designated as *inversio testis* and *ectopia testis*. By inversio testis is understood that condition in which the testicle is rotated upon its own axis; by ectopia testis we mean the condition in which after its passage through the inguinal canal, the testicle is found under the skin of the abdomen, or beneath the crural arch, etc. The most important variety of this anomaly is ectopia perinealis. In this form the testicles lie to the right or left of the median raphe between the anus and the scrotum. They are generally atrophic and movable, but can never be pushed into the scrotum. It is caused by defective size of the scrotal partition. Both inversion and ectopia are often combined with hernia.

ACUTE ORCHITIS AND EPIDIDYMITIS

These may result from trauma, from gonorrhœa, or they may follow a series of infectious diseases. The connection between orchitis and epidemic parotitis in particular was known even to Hippocrates. The disorder is far more frequent in adults than in children, and is dependent, according to Soltmann, with a difference in the chemical composition of the tissue of the testicle. Some authors regard the trouble as a sympathetic one, others believe it to be due to metastasis. It is said to manifest itself between the third and eighth days of the primary disease. According to Steiner, the infectious agent of parotitis may produce "mumps" of the testicle, skipping the parotid gland, and vice versa.

Symptoms.—The affection is attended by redness, swelling, and severe pain.

The **treatment** is symptomatic and consists in elevation of the part, and the use of the ice-bag. Atrophy of the testicle may result. The statement that involvement of the genital organs is more common in boys than in girls is said to be only apparently true, because the disorders of the ovaries are of a more vague and indeterminate type, and they are not so accessible to examination. Of the other infectious diseases which may produce inflammation of the testicles, must be mentioned varicella, variola, and scarlet fever. Spolverini described gangrene of the testicle following an inflammation of the testicle due to the bacterium coli communis. Traumatic epididymitis due to contusion, may according to Griffith, be the cause of general convulsions in children.

SYPHILIS AND TUBERCULOSIS OF THE TESTICLES

According to Hutinel, syphilitic changes in the testicles are to be found in one third of all syphilitic infants. They are exhibited as interstitial orchitis and epididymitis, and are later, perhaps, a cause of sterility.

Tuberculosis of the testicle is most common in the first two years of life, according to Broca, who saw 11 cases among 16,600 children who were admitted to his service. The affection shows some striking differences from the condition in the adult. In children the onset is often acute with the formation of a large tumor, which may soon subside. Both the general and the local prognosis is said to be better in children than in adults. Contrary to the statements of other writers, Broca never saw the trouble spread to the glands of the peritoneum and mediastinum. In babies there is a tendency to early abscess formation, but without the formation of fistulæ, which are less common in infancy than in adult life.

TUMORS

According to Steffen, who collected 19 cases, malignant tumors of the male genital organs are rare in infancy. Of the 19 cases, six involved the prostate and 13 the testicles. Cancerous degeneration of the prostate has hitherto been observed only in children up to eight years of age; the youngest was six months old. The tumor is generally a medullary carcinoma. It may attain considerable size, and may produce symptoms due to displacement of the pelvic organs. Metastasis has been found in almost all of the internal organs, but it is not certain whether in adults it affects the bones. Pain and difficulty in urination are the earliest symptoms. A point of importance from the standpoint of diagnosis is that the neighboring inguinal glands are early involved. The general health soon suffers. The diagnosis is established by palpation through the rectum. The fatal termination, which as a rule occurs in from three to seven months, cannot be averted even by operative interference.

Cancer of the testicles, somewhat more frequent than cancer of the prostate, attacks by preference the very youngest children, often during the first few months of life. The youngest case of the kind was only six weeks old. Medullary carcinoma and scirrhous forms have been described. The former are both more rapid in growth and more malignant.

At the onset, cancer of the testicle is not attended by any symptoms. It seldom causes pain. As it increases in size it causes discomfort by its weight. The general health is noticeably impaired, and a decided cachexia appears. The prognosis is unfavorable because of the great tendency to metastasis. Only an early operation can promise any hope of a cure. Rarer than carcinoma are the sarcomata. Amongst the benign tumors are to be mentioned enchondromata and embryomata.

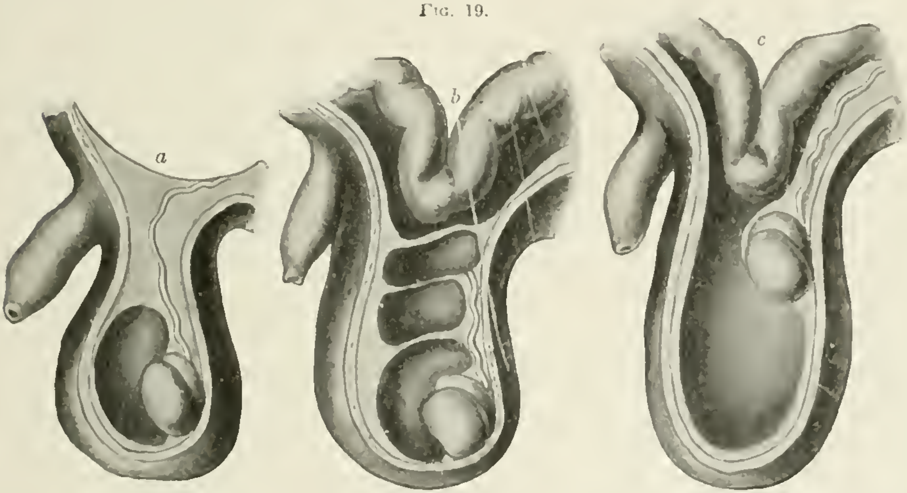
HYDROCELE—HÆMATOCELE

A double serous sac envelopes the testicle and the epididymis, which under normal conditions contains only a few drops of fluid. An increase in the quantity of fluid distends the sac (hydrocele) and enlarges the scrotum. If the distention is caused by blood it is called a hæmatocele.

Hofmokl gives the following division of the disorder, which may be either congenital or acquired:

1. *Hydrocele vaginalis testis et spermatici* (congenital).
2. *Hydrocele vaginalis funiculi spermatici communicans* (congenital and acquired).
3. *Hydrocele in combination with a hernia* (hydrocele herniosa).
4. *Hydrocele vaginalis funiculi spermatici uni- et multilocularis* (non-communicating).
5. *Hydrocele vaginalis testis simplex.*
6. *The biloculary form of Kocher,* of which the location is in the distended extremity of the vaginal sac, partly in the peritoneum, partly in front of the external crural ring.

Fig. 19.



a.—Common form of hydrocele testis. The tunica vaginalis propria is extended by fluid, and the parietal peritoneum runs smoothly over it. *b.*—Hydrocele testis, hydrocele funiculi spermatici, and inguinal hernia. The processus vagin. periton. has grown together in several places, and has in consequence formed several sacs lying one above the other. At the bottom of the scrotum is a hydrocele of the testis, above it two hydroceles of the spermatic cord, met above by the hernial sac. *c.*—Hydrocele communicans, secondary inguinal hernia. In consequence of an incomplete descent of the testicle, the union of the processus vaginalis is defective. A hydrocele communicans has developed, which is simultaneously the seat of a hernia. This condition is also called hydrocele hernialis.

Kocher, Cohnheim, Birch-Hirschfeld consider the hydrocele as the product of an inflammatory irritation of the membrane of the testicle or of the spermatic cord. Kocher therefore proposes the title chronic serous periorchitis or chronic serous perispermatitis. Intra-uterine inflammation of the testicles and spermatic cord are made responsible for the congenital form. Wechselmann denies the supposition that it may be due to circulatory disturbance.

The **symptomatology** is the same as in adults, but the diagnosis is easier owing to the thinness and transparency of the tisuses.

For the **differential diagnosis** from herniæ and inguinal testicles we refer to works upon Surgery. *Expectant treatment* is justified because in the course of weeks or months spontaneous recession occurs in many cases. If this does not take place, aspiration, with or without the injection of a few drops of iodine, may be tried.

Hæmatocele is very rare in children, although they may result from contusion during delivery in the breech position.

DISEASES OF THE FEMALE GENITALS

ANOMALIES OF FORMATION AND POSITION

Fissure of the urethra and clitoris has been observed in females corresponding to epispadias in the male.

Defective opening of the urethra has been observed in various locations, as for instance in the intestines, or into the vagina.

The urethra may be partially or entirely missing, and then the bladder opens directly into the vagina.

If a fissure exists in the vaginal urethra we speak of it as a hypospadias. Bitner and Mosenthal designate as hypospadias the opening of the urethra upon the vestibule as a blind sulcus, while the true vaginal urethra opens upon the anterior wall of the vagina either immediately behind the hymen or further back.

It speaks for the rarity of hypospadias in the female sex that Hofmokl in 104,446 children did not once find this deformity in girls, while he found 13 instances in boys.

Among the atresias of the external genitals are to be mentioned: Complete atresia of the anus and vulva, and atresia of the anus and vagina. The atresia of the vulva may consist simply in an epithelial adhesion of the opposing surfaces of the labia majora and minora, or in a solid union of the parts. The result of atresia of the hymen in the newborn is the retention of mucus, in girls after puberty a hæmatocolpos.

Congenital cysts of the hymen, described by Ziegenspeck, are also due to defects of development.

Among the deformities of the ovaries are to be mentioned: defective formation and accessory glands. Defects of formation of the uterus and vagina are due to the manner in which the Müllerian ducts become united into a single structure.

Where the development of the uterus and tubes is defective there is free mobility of these organs, and a disposition to hernia of the ovary, which occurs not very rarely. Prolapse of the uterus which is seen in newborn children, is also to be referred to congenital anomalies. The diagnosis of congenital anomalies, according to Zuppinger, is very difficult, and must be made with great care, particularly if an operation is contemplated.

HÆMORRHAGES FROM THE GENITALS

Schulkowski observed in 10,000 newborn children (girls), 35 cases of hæmorrhage from the genitals. The metrorrhœa, usually extremely scanty, and seldom abundant, never appeared before the fifth and generally at the sixth day. Schulkowski refutes the contention that in these cases the hæmorrhage was a precocious menstruation, as some writers believe. Neither does he believe the hæmorrhage can be ascribed to asphyxia, breech presentation, or instrumental delivery. He sees the cause as a physiological hyperæmia of all the abdominal organs which is present after birth. Zappert believes, as a result of histological examinations, that the cause of the hæmorrhages is a physiological irritation of the uterine mucous membrane. Henoch connects the vaginal hæmorrhage with the shedding of epithelium that appears after birth, and designates as further causes papilloma of the vulva and vagina. He calls our attention to the fact that vaginal hæmorrhage may be imitated by bleeding from the polypoid prolapse of the urethral mucous membrane, which in cases of great severity may be represented as a dark red protrusion between the walls of the genital fissure.

Precocious menstruation, is in any case, extremely rare as a cause of hæmorrhage. It is accompanied by other evidences of general maturity. In older girls hæmorrhages are described which may be due to hypertrophy of the cervix, fungous endometritis, salpingo-oöphoritis, metritis (of gonorrhœal origin), cystic degeneration of the ovaries, or to causes belonging to diseases outside the genital apparatus entirely.

It seems useless to treat the hæmorrhages of the newborn which are due to physiological irritation, since they cease in a short time of their own accord. The treatment of the other forms is based upon the etiology.

TUBERCULOSIS AND TUMORS

Tuberculosis of the female genital organs is generally a secondary affection in which the tuberculous virus reaches the peritoneum first, and from there spreads to the Fallopian tubes or else it reaches the intestines and then the vagina and peritoneum, and finally involves the tubes. According to Brüning, who collected 40 cases, the tubes generally show a caseous alteration, and then the uterus, vagina, ovaries, and vulva. He is of the opinion that primary genital tuberculosis, as described by Demme, Schlenk, v. Karjahn, and others, has not yet been proven to occur, because the diagnosis was only established clinically. Vierordt in one case established the diagnosis by finding tubercle bacilli in the discharge. It may be impossible to establish the diagnosis when symptoms of general peritoneal infection are present.

Among the *benign tumors* are to be mentioned in the first place, the vaginal polypi which are characterized by hæmorrhages. *Malignant tumors* of the genital organs are found more often in girls than in boys. The majority of the cases are seen in the first six years, according to Steffen, who collected 33 cases from the literature. The favorite seat is the vagina, where 15 out of the 33 tumors collected by Steffen, were located. The uterus was affected in 4 cases. In 9 cases malignant tumors of the ovaries were found, a condition to which the age of puberty seems to have a predisposing influence. The majority of the tumors observed during the early years of life seemed to be congenital.

Sarcoma of the vagina and its variations are amongst the most important forms. According to Pick, who observed 15 cases of polypoid vaginal sarcoma, they have a tendency to spread with great rapidity, to intrude into the neighboring tissues, and to infiltrate them with cancerous degeneration. Before they are visible, the symptoms are profuse hæmorrhage, disturbances in micturition, and pains in the abdominal region. Even a very early operation will not protect the patient from recurrence.

The uterus may be affected by both sarcoma and carcinoma. The symptoms are the same as those of the same affections in adults. The malignant tumors of the ovaries are generally carcinoma. The diagnosis cannot often be confused with malignant tumor of the abdomen. The prognosis, in spite of operative interference, is bad owing to the tendency to recurrence.

VULVOVAGINITIS

Practically the most important disease of the female genital apparatus is the inflammation of the mucous membrane of the vulva and vagina. It is only in the last two decades that we have come to realize their terrible frequency. But the experience of the last few years has also taught us that the great majority of the cases are infectious, and are due to the same organism that causes the urogenital gonorrhœa of the adult: namely, the gonococcus of Neisser. In the following remarks therefore we shall speak of the gonorrhœa of girls, and only incidentally will mention be made of inflammations due to other causes, and of their terminations.

(*a*) GONORRHŒAL VULVOVAGINITIS

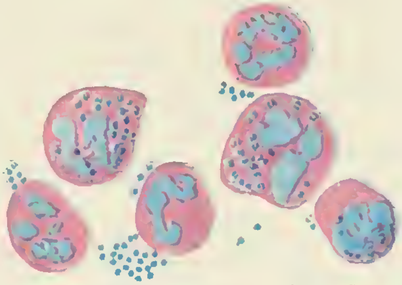
This affection, according to Pott, is to found most frequently in the newborn and during the first five years. Later on it increases again in frequency at the age of puberty. In Pott's statistics there were 3,921 girls, of whom 44 were infected with the gonococci, and of these 27 were less than five years old. Epstein does not believe that there is any special predisposition between the second and fourth years, but that in numerous cases brought to the physician for examination the infection occurred at the very beginning of life, and only attracted attention when the child began to walk.

Etiology.—The exciting cause is the gonococcus discovered by Neisser. Even during delivery the infection may enter the child's genitals from the vagina of the mother affected by gonorrhœa, but later on in life, too, there are a whole chain of circumstances which may render the infection possible. For instance, sleeping with the infected mother in the same bed; the use of the same wash-basin, the same chamber, the same bath water, the same towels; contact with hands soiled by the gonorrhœal poison; taking the temperature with a thermometer dirtied by a previous record. Moreover, especially in large cities, coitus practiced from perversity or for superstitious reasons, is a deplorable cause of the disease.

In this place it is necessary to consider why it is that more girls than boys become infected. Poet explains this difference by the fact that in boys owing to the firm adhesion of the prepuce to the glans, the mouth of the urethra, funnel shaped as it is, is so small that it will only permit the entrance of a very small sound, whereas in girls the genital orifice is free and open.

Symptomatology.—In many cases the infection will not be betrayed by a single subjective symptom. The mother as a rule brings the child to the doctor because the clothing is stained with yellow spots. Examination shows the vaginal orifice to be more or less irritated, swollen and reddened, and in more advanced cases there are found small ulcerations and erosions upon the labia majora and minora. From the vulva a discharge escapes which is sometimes chiefly mucous, sometimes chiefly purulent. When stained with aniline dye, we find under the microscope, in addition to leucocytes, great numbers of typically located gonococci (Fig. 20).

FIG. 20.



Pus from a vulvovaginal gonorrhœa. Stained according to May-Grünwald.

If there are subjective symptoms, they present themselves in the form of dysuria, itching of the genitals, associated with more or less extensive eczema, due to the secretion, and eventually considerable pain in walking. In cases of long duration the general health suffers, the appetite fails, the temper is irritable, and the child loses its freshness and becomes pale. An elevation of temperature is not rare.

Diagnosis.—The most striking thing is the extraordinary ease with which the gonococci can be demonstrated. Their form, intracellular location, and the negative result of Gram's method permit them to be recognized without even the necessity of a culture.

Sometimes upon inspection are seen injuries of the genitals which point to the commission of a "coitus." Unless there are evidences of violence one must be exceedingly cautious in surmising a crime, since there are many other possible ways of infection.

Course and Complications.—The course of the disease is eminently chronic. Only under unusually favorable circumstances will the disease disappear in six weeks. Generally it lasts considerably longer, even for years, during which latent periods alternate with periods of acute exacerbation. The complications consist in the first place in an extensive participation of the genito-urinary mucous membrane in the inflammatory process. In this way there will develop inflammation of the membrane of the urethra and bladder, which will be attended by severe urinary distress. Less frequently an infection of the cervical and corporeal uterine membrane results, which may lead to hæmorrhage. Bartholinitis is rare, as is the spreading of the gonorrhœa to the peritoneum, the gonorrhœal peritonitis which may lead to death by sepsis and pyæmia, but in which there have also been a few recoveries.

Among the more common complications is *gonorrhœal arthritis*, which most often affects the wrist and tarsus, and less frequently the jaw-, finger-, and toe-joints. The symptoms are pain and swelling of the joints. In addition we may see tendovaginitis, and an extremely rare gonorrhœal stomatitis. Paulsen has described a gonorrhœal exanthem, with the formation of blisters and pustules, the occurrence of which is said not to be so very rare.

The **prognosis** is serious in so far as the disease is persistent and frequently relapses. Even though it be true that a cure is generally effected after some months, it must not be forgotten in our estimate of the prognosis that death sometimes occurs from the complications, and that the disorder may be accompanied by diseases of the genital organs which may show themselves only at the age of puberty (endometritis, etc.).

Prophylaxis.—The readiness with which girls may be infected by the gonococcus indicates that the prophylaxis should attempt in every possible way to prevent exposure. Epstein proposes that the vulva should be carefully cleansed in the first bath given after birth, and that, if there is any suspicion of gonorrhœa in the mother, there be allowed to trickle into the vagina a 2 per cent. solution of silver. In addition all who nurse the mother and child should be instructed to attend to the child first and the mother afterwards, and that they observe at all times the most rigorous antisepsis, just as in a surgical operation. If we wish to avoid all danger of spreading the infection, the utmost cleanliness, the most careful antisepsis, and the strict prohibition of the use of the same towels, crockery, or sponges are absolutely necessary. Especially in hospitals, infirmaries, and boarding schools, where many girls sleep together, are the most energetic precautions necessary. Koplik is right in demanding that every newly admitted child should be submitted to a bacteriological examination of the secretion of the vulva and vagina, in order to know the possible origin of infection, and if necessary to isolate the case at once.

Treatment.—This consists in repeated and thorough cleansing of the genitals. Once or twice a day a sitz bath in a 1–1000 solution of tannic acid is given. As long as there are still more or less acute symptoms, it is possible to effect a cure in the course of one or two months by these means, combined with rest in bed, alone. It is questionable whether irrigation of the vagina, which is proposed by some, is of great benefit or cures the disease any sooner. And it may be mentioned that injections in the vagina often cause severe pain to some of the children, and moreover, it is apt to turn the child's attention to the genital apparatus, which is very undesirable from a pedagogical standpoint. For use with the syringe there have been proposed a 0.5 per cent. solution of protargol, a 0.5 per cent. solution of sublimate, a rose red premanganate solution. The injection of the vagina should only be done by the physician himself, and he should employ a syringe protected by a soft rubber tip. After the injection a tampon or small rod of iodoform may be introduced into the vagina. Perhaps a still better thing is the introduction of a tampon saturated with a 10 per cent. solution of ichthyol, which is squeezed out into the vagina. Small doses of sandal oil may be given internally. It is necessary to protect the eyes against the possibility of infection. Where there is inflammation of the upper genital tracts, the strictest rest in bed is imperative. Gonorrhœal arthritis is probably best treated by Bier's method.

(*b*) VULVOVAGINITIS OF OTHER ETIOLOGY

Epstein describes a catarrh of the female genitals in the newborn which is part of a desquamation process, which occurs in all the mucous membranes and upon their apertures. This catarrh is present in fœtal life and continues energetically after birth in consequence of the changed conditions of life and the new external impressions which affect the newborn. The catarrhal process is demonstrated by an abundant secretion from the vulva. In the beginning it contains viscous hyaline masses, but later on these may become more liquid as a result of the increase of microörganisms, and the secretion may assume the aspect of a blennorrhœa. This process which Epstein calls desquamation catarrh may be accompanied by a catarrhal vulvovaginitis from inattention to cleanliness of the genitals.

In the secretion are found epithelium, leucocytes, and microörganisms of the most varied types, but never the gonococci, which latter fact enables us to make the diagnosis.

Von Hansemann describes a very early form of catarrh of the female genitals due to spreading of morphological particles from the vagina of the mother. This catarrh never becomes chronic.

Aphthous vulvitis is the name given by Parrot to an affection, occurring in infants, which consists in the formation of circular white and grayish white plaques, from 1–4 mm. in size, on the labia majora and minora, but which may spread to the adjacent parts in the median line or to the posterior portion of the genital region, and sometimes to the genitocrural and inguinal folds. After from two to three days there appear elevations with yellowish coating and a red areola, which cause itching and swelling of the surrounding parts. A serious but very rare complication is gangrene. The disorder seems especially prone to develop after the infectious diseases, particularly measles, and less frequently it occurs idiopathically. Dusting with iodoform promotes healing and prevents gangrene.

An *impetiginous form of vulvitis* is described, characterized by the formation of blisters and pustules, beginning with redness, and with a reactive inflammation and catarrh. This affection is an unusual localization of the common impetigo contagiosa. In making the differential diagnosis we have to exclude syphilis, herpes, and gonorrhœa. Treatment consists in the use of 10 per cent. solution of hydrogen peroxide.

Vaginal catarrh may also result from *herpes* and from the localization of *syphilitic lesions*, and from *vaccine pustules.* The diagnosis is generally to be made by a careful examination. Furthermore there appears in scarlet fever, in measles, and in smallpox during the acute stage, a vulvitis, which Henoch considers to be an extension of the skin inflammation over the vulva. General dyscrasiæ are also regarded as causes of vulvovaginitis in infancy, and we speak sometimes of "vulvovaginitis cachectica and scrofulosa."

It is hardly necessary to emphasize the fact that infection due to foreign bodies introduced into the vagina, may also produce a vulvovaginitis.

In comparison with gonorrhœa, the above causes of inflammation of the genitals are rare. Cleanliness and antiseptic washes will effect a cure in most cases.

There still remains to be mentioned a disagreeable complication of the various forms of inflammation which we have been describing, and that is the phlegmon of the vulva and vagina, which is attended by considerable swelling, severe pain, œdema of the surrounding parts, and serious febrile reaction.

Treatment consists in poultices of acetic alum, sitz baths, and the early evacuation of the pus. In the most serious cases, gangrene of the labia with the formation of violet colored, rapidly softening, and ill smelling spots, may develop from the phlegmon, or from an added erysipelas. This gangrene may appear apparently primarily, without any phlegmon, in the course of infectious diseases, which are attended by a marked cachexia. Henoch compares it to the noma of the cheek. Treatment consists in early incisions and the bold use of the cautery. The general strength must be maintained by a concentrated diet and stimulants.

THE PECULIARITIES OF THE CHILD'S NERVOUS SYSTEM

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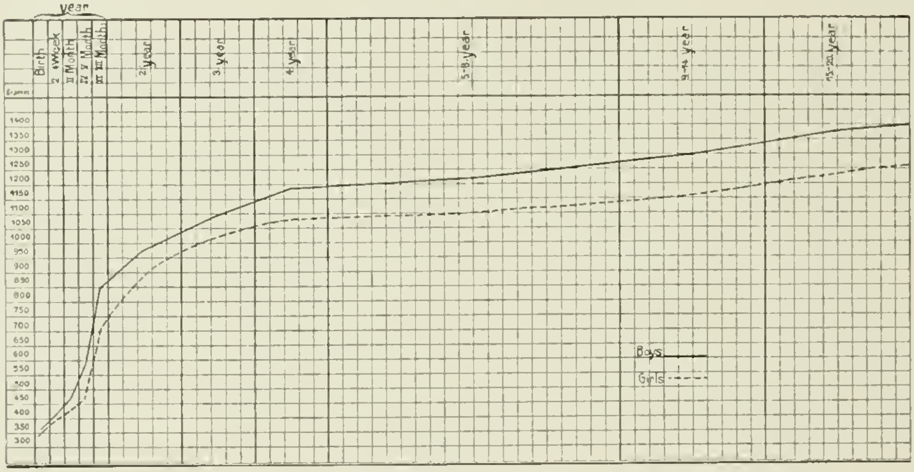
MORE than most of the other organs the central nervous system of the newborn and growing child exhibits peculiarities which serve to distinguish it from that of the adult. We have not merely to do (as in many of the other organs) with a structure which is still immature but functionally active, and which simply goes on to full development, in direct proportion to the general growth of the body and with only slight changes in its external configuration. The central nervous system of the newborn and even that of the older child, is not simply quantitatively, but it is also and more especially qualitatively, an inadequate organ, which at the outset, moreover, is physiologically active only in part. Substantial alterations must therefore take place before it acquires the significance it possesses in the later phases of the individual's life history.

It is impossible to give in the brief space alloted to us, an exhaustive review of the anatomic and functional differences which exist between the nervous system of the child and that of the adult. Only the more important peculiarities bearing upon the gross and minute morphology of the central nervous organs will be considered; their significance from the standpoint of psycho-physiology being at the same time discussed. The facts to be deduced from this study will amply demonstrate to the physician, how eminently important, not only for the child's immediate welfare, but through its far-reaching influence, for the entire course of the individual's existence as well, is the hygiene of infancy and early childhood, and the proper management of whatever nervous disturbances may arise at this early period of life.

MORPHOLOGIC PECULIARITIES

Aside from the intimate adhesions between the dura and the bones of the cranial vault, the absence of Pacchionian bodies and the smallness and friability of all the structures, the mere opening of the skull and vertebral canal presents in children no peculiarity worthy of note. The disposition and form of the various structures, their relations to the surrounding parts, are the same as in the adult. Only in the vertebral canal are slight differences to be observed. It is well known that the adult spinal cord terminates at a point corresponding to about the lower third of the first, or to the superior border of the second, lumbar vertebra. In the fœtus, on the contrary, the cord fills the vertebral canal entirely and it is only in the later stages of intra-uterine life and owing to the more rapid growth of the vertebral column, that it gradually recedes. In the newborn, the end of the conus medullaris still lies in the cavity of the third lumbar vertebra; the ultimate relations however, become more or less fully established during the first phase of life. This modification in the relations between the spinal cord and the vertebral column affords an explanation of the fact, so significant to the surgeon, that in young children and particularly in infants, the spinal nerves, especially those of the lower segments, pursue, in order to reach their respective foramina, a different course than in the adult, being much less obliquely directed.

Fig. 21.



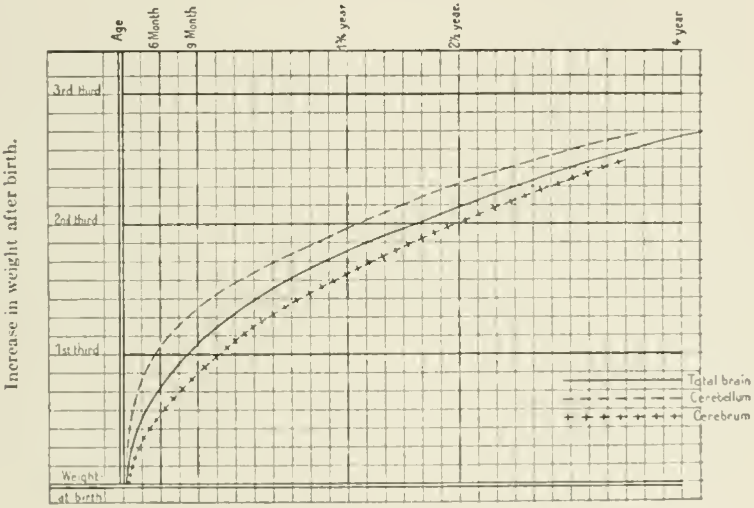
Average growth of the brain in boys and girls. The periods of time after the eighth year are shortened in the chart.

On section, the cut surface of the central nervous organs more particularly of the prematurely-born, has a very distinct grayish hue which is not to be seen in the brains of older individuals.

The *brain* of the newborn is relatively large. Whereas most of the organs of the body weigh, at birth, only the tenth to the fourteenth part of their ultimate weight, the brain has already reached the fourth part of its ultimate weight. In proportion also to total body weight is its volume greater in the infant than in the adult; the proportion being as 1:85 and even 1:75 according to Ziehen's estimates, against 1:42 in the male, and 1:40 in the female, adult. Mies places the absolute weight of the brain of the newborn at about 340 grams in males and 330 grams in females. These figures, he obtained from statisties published prior to 1894, and based on single weighings of brains of the most miscellaneous source (brains of Germans, Romans, Slavs, as well as of subjects belonging to remote countries), having thus used an anthropologic material of an insufficiently equivalent and uniform character. Ziehen, Marchand and others, consider that these figures are rather too low.

It must certainly be stated that among healthy Germans the average *weight* of the brain of the newborn is decidedly higher (between 350 and 370 grams). With the progress of development, the weight of the brain steadily increases and may even reach 1400 grams; the average figure varying between 1250 and 1275 grams. According to Mies, the maximum figure would be only 1230 grams, and 900 grams would represent the average weight of a fairly well developed brain. The first third of this increase in the weight of the brain had taken place (in the author's cases) by the ninth month, the second third by the second quarter of the third year, of extra-uterine life (see Figs. 21 and 22); thus, the original weight of the brain was doubled by the end of the third quarter of the first year and trebled before the expiration of the third year (Marchand). According to this author, the ultimate weight of the brain is attained between the nineteenth and twentieth year.

Fig. 22.



Growth of the brain in the first four years.

At all ages the average weight of the brain is lower in females than in males. In consequence of the relatively more vigorous development of the male brain, this difference in brain weight between the sexes, which, at birth, only equals from 10 to 15 grams, increases to 120 grams and more in adult life.

Differences in brain weight also occur which are not dependent upon corresponding differences in total body weight or general development, as they are observed in subjects belonging to the same sex, of the same age and even of the same weight. They are to be considered simply as the expression of some particular individual (most often hereditary) tendency. Even in the brains of the newborn, differences of from 50 to 70 grams may be observed, as also, variations in the capacity of the cranial cavity, which, already in the third week, may amount to as much as 75 and 100 c.c., and even more. As age advances the scope of this physiologic variation becomes even greater. In children of about three months, differences in brain weight of from 200 to 300 grams have been recorded. Very frequently individuality finds expression in a precocious development of the brain. Brains weighing as much as 1280 grams have been observed in children of only three years, and from 1350 to 1400 grams, in boys of five years. Excessively high figures have also been occasionally recorded; in Lorey's case (a child of six years) the brain weighed 1840 grams.

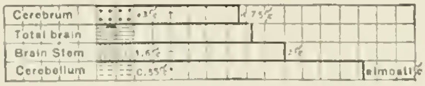
The weight of the brain proper (cerebrum) has, at birth, a minimum figure of from 305 to 320 grams, a maximum figure of from 310 to 345 grams. The extra-uterine gain in weight may, in a general way, be placed at from 830 to 840 grams, reaching in certain instances however, as high as 910 grams. At all ages, the average weight of the male brain is greater than that of the female. It is the weight of the cerebrum that heredity more particularly influences.

Next to weight, the *dimensions* of the child's brain give us an insight into the pronounced metamorphosis which takes place after birth. From the original measurements of 9 cm. in the fronto-occipital pole-length, 7 cm. in width and 5 cm. in height, does the brain acquire, often enough even before the time of puberty, the ultimate dimensions of 16–18, 13–15, 8–9 cm. respectively.

A feature which is very distinctive of the cerebrum of children, and more particularly of that of the newborn, is the relatively poor development of certain of its parts. Thus, for instance, the frontal lobe, especially in the prematurely-born, is decidedly inconspicuous. The insula of Reil, though its external markings may be appreciable, is not as distinctly outlined in the first months of life as it is later.

In connection also with the fissures of the brain, are peculiarities to be noted in the new and prematurely-born. The primary fissures are, in proportion to the general development of the organ, relatively deeper during the first quarter of life than later. It is probable that as long as the phase of rapid growth determined by the investment of nerve-fibres with myelin sheaths lasts, further changes take place in connection with the shallower fissures of the convexity, such as confluence of superficial sulci (which, in the parieto-occipital region, occur with especial frequency during the first weeks of life), or deepening of fissures owing in the latter case, to the progressively increasing prominence of certain convolutions.

The *cerebellum* of the child also presents certain points of interest. From about 18 to 21 grams at birth, its weight increases in the course of development to 120 or 130, and even to 135 and 150 grams. The first two-thirds of this increase in the weight of the cerebellum takes place much sooner than the corresponding increase in the weight of the cerebrum or of the brain generally, being usually manifest anywhere from the sixth month to the end of the second year; whereas its ultimate weight. corresponding to the very slow progress of its further growth, rarely obtains before the end of the second decade. The average weight of the female cerebellum is invariably less than that of the male. This difference between the sexes increases from about three grams at birth, to about 15 or 20 grams in adult life. Independently of total body weight or general development, and in subjects of the same age, by no means trivial variations may be observed in the size of the cerebellum, ranging all the way from 10 grams during the first months of life to 30 grams and more later. As early as the eleventh or twelfth year, the weight of the cerebellum may equal or exceed that of the fully matured organ; which circumstance, however, does not necessarily imply a corresponding increase in the total weight of the encephalon.



Comparison of extra-uterine increase in size of the brain and its component parts from birth. The figures give the weight of the parts of the brain in comparison with the entire brain at birth and at the end of brain growth.

The *brain stem* (medulla, pons and quadrigeminal region) increases in weight from about 5.5 grams at birth to about 27 or 28 grams, in direct proportion to the growth of the brain in general. It is slightly larger in males than in females.

From the foregoing facts, it follows that the brain as a whole in the course of development quadruples its original weight at birth; the cerebrum likewise, almost as much; whereas the brain stem increases to five, and the cerebellum to seven times its original weight. Thus, is the development of the cerebellum not only more rapid, as has already been shown, but, in proportion to its original weight, also much more considerable, than that of the other parts of the brain. For this reason, the relative proportion between the size of the various segments of the brain is subject to constant variation: the cerebrum sinks from almost 93 per cent. of the total brain weight (at birth) to 87.5 per cent.; the brain stem rises from 1.6 per cent. to 2 per cent. and the cerebellum from 5.5 per cent. to almost 11 per cent. of the total weight.

As regards the *spinal cord*, considered as beginning at the inferior limit of the pyramidal decussation (and deprived of its nerve roots and dura mater), its average weight in the newborn varies between 3.0 and 3.4 grams, and between 27 and 28 grams in adults. Its weight increases rapidly at the beginning of extra-uterine life, being doubled by the fifth month, trebled at the end of the first year and quadrupled at the beginning of the third year. Its average weight is practically always greater in males than in females. The relative proportion between the weight of the spinal cord and that of the brain also differs in children from that in adults; being in the latter about as 1:50. In the newborn this proportion equals 1:110 at the very most; it becomes 1:80 in the third year and reaches 1:67 by the end of the sixth year. Thus, with advancing age, the weight of the spinal cord becomes ever greater, relatively, than that of the brain; whereby it necessarily remains relatively less in males than in females of the same age. In children of the same age and sex, a heavy brain generally implies a proportionately heavy spinal cord.

	At Birth		1 Month		2–3 Months		4–6 Months		7–10 Months		11–15 Months		16–21 Months		22–36 Months		37–56 Months		57–91 Months		11⅓ Years		14¼ Years	
	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀	♂	♀
Total Brain	(370)	(350)	431	396	492	471	623	576	758	692	872	820	1018	891	1100	1046	1152	♀ 1067	1273		1290	1252	1301	
Cerebrum	(340)	(320)	409	368	452	439	558	517	664	616	773	698	890	753	967	911	1003	939	1006	(1071)	1121	(1102)	1140	
Cerebellum	21	18	28	24	36	35	51	(51)	77	67	90	84	112	101	122	117	130	♀ 122	135		136	(128)	139	
Brain Stem	5.5		9.0	7.9	11.2	10.7	12.7	11.8	11.2	13.4	14.7	14.4	16.5	16.0	17.5	16.8	19.0	18.3	20	19	♂ and ♀ 21			
Spinal Cord	(3.4)	(3)	3.9	3.8	5.0	4.6	7.1	6.1	8.2	7.5	10.7	(10.5)		(11.0)	(13.0)	13.6	15.7	14.8	18.9	(18.2)				

Average weight of the total brain, of single divisions and of the spinal cord in children.

The length of the cord at birth equals 14 cm., and only at the beginning of the fifth year is an increase of about 10 cm. appreciable; it very slowly reaches its definite length of 43–45 cm. Whereas at birth, it corresponds to about 29.5 per cent. of the total body length, it steadily looses ground after the end of the first year, ultimately representing only 25 to 26 per cent. of the same. The configuration of the spinal cord also undergoes considerable modification, especially at the level of the enlargements, where, in the newborn, and not infrequently for some time afterward, the inequality of the two diameters on cross-section is scarcely appreciable.

Let us now briefly outline the *histologic peculiarities* of the child's nervous system. Developmental processes characterized by an increase in size and number of the various constituent elements take place throughout the central organs, and gradually, the complex structures formed by the agglomeration of membranes, blood vessels, nerve fibres, etc., appear. The luxuriant growth of the cellular elements (cell and nuclear division) especially at the very beginning of life, the streaming out of axis cylinders and blood vessels, the splitting off of glia fibres, realize, all taken together, a histologic aspect, which has been designated not wholly without reason, as the nearest approach to true inflammatory reaction. As regards nervous tissue proper, it is to be noted that in the newborn, the ganglionic cells do not present everywhere the specific characters of the adult type of cell; this being especially true of those of the cerebral hemispheres. In part, they remain dense of texture and present, in both nucleus and cell-body, structural peculiarities (Arborio) or an embryonal character; the characteristic configuration more particularly of the pyramidal cells is still wanting. Nuclear division is to be seen in cells undergoing developmental changes; being especially common at the very outset and becoming ever rarer in subsequent phases of development. Pigment is totally wanting at birth, and only at a later period does it very gradually appear. The dark brown pigment of the locus cœruleus appears at about the end of the first year; that of the nucleus of the vagus and substantia nigra, in progressively increasing quantity after the third or fourth year; the light yellow pigment of the posterior spinal ganglia is rarely found before the sixth, while that of the spinal cord only appears after the seventh or eighth year. In childhood the cells of the cerebral hemispheres are devoid of pigment.

It is in connection with the great mass of *nerve fibres* of the central organs, however, that the most marked changes are observed. Sprouting of nerve fibres (differentiation of new fibrils) takes place throughout the central nervous system from the very beginning of life. The extra-uterine increase in volume of its various segments is due, above all, to the fact that great numbers of nerve fibres become invested with myelin sheaths only after birth; that being especially true of those of the cerebral hemispheres. Thus, while the spinal cord possesses, at birth, practically the full measure of its myelin constituent, on the contrary, the brain stem and cerebellum, with the greater part of their substance, and extensive areas of the cerebrum are totally devoid of it; a fact which explains the grayish hue observed over large portions of a freshly cut surface of this organ as was previously stated. In children born at the eighth or ninth month, practically the only tracts of fibres which are provided with myelin sheaths, are those subservient to general sensibility, tactile and muscle-sense; at a later period, the pyramidal tracts may also be partly invested. In children born at full term, in addition to the above-named tracts, the fibres belonging to the olfactory and visual systems, as well as to certain segments of the corona radiata, also possess their myelin sheaths in greater or lesser numbers; whereas considerable portions of the temporal, occipital and frontal lobes, as well as of the commissural system are still practically wholly unprovided. The researches of Flechsig, Siemerling, v. Monakow, Probst and others, have shown that the various tracts of fibres are not affected simultaneously by the process of myelinization, the individual fasciculi receiving their sheaths at different periods. We know however, that the constituent fibres of tracts belonging to the same system, or to systems associated in physiologic activity, do become invested with myelin at the same time. Only after the ninth month of extra-uterine life is the projection system of fibres fully provided with myelin. At birth, the cerebral cortex contains but very few tangential fibres widely scattered over its entire surface; it is only in the course of many years that these intracortical tangential fibres, as well as the numerous and larger subcortical association tracts, acquire their full development. Whether these various fasciculi, once formed, retain their original structural proportions, or whether they are influenced by the varying degree of functional activity, so that new axis cylinders and myelin sheaths develop in numbers adequate to the exigencies of acquired physiology, as Edinger has suggested, the problem is hardly solvable, owing to the appreciable variations encountered in the histologic configuration of these same tracts.

The very considerable alterations which occur in the central nervous system in the course of development, affect not only its ganglionic cells and their axis cylinder processes, but its supporting framework or neuroglia tissue as well; being especially active in the very first phase of extra-uterine life and most marked on the surface of the brain and in the ependymal structures. Increase in number and differentiation of cellular elements, structural reorganization of fibrillar networks, take place, affecting especially the central gray matter which surrounds the Sylvian aqueduct and the central canal of the spinal cord. Both these structures are relatively wide and gaping in infancy and early childhood; a condition which is particularly favorable to the free circulation of bacteria and toxins over considerable portions of the important columns of gray matter which constitute their limiting walls.

PHYSIOLOGIC AND PSYCHOLOGIC PECULIARITIES

The distinct features just described in connection with the morphology of the child's nervous system let it at once be supposed that its function, likewise, is still imperfect and atypical. Indeed this fact already becomes manifest on examination of the peripheral nervous system. The excitability both of motor nerves and of muscles, tested by means of faradic and galvanic electricity, is decidedly faint until about the eighth week; the muscular response is sluggish (C. Westphal, Soltmann, A. Westphal). Altogether by degrees the characteristic features of the adult response become well-defined, and then only after a long phase of hyper-excitability (the so-called spasmophile period), which, by some, is regarded as normal, but which Mann more correctly considers pathologic, and the indication of a widespread diseased state, however insignificant in its intensity. The sensory nerves also show but slight sensitiveness to electricity for some little time after birth; in the newborn, the face itself is totally insensitive to even strong electric currents.

The very considerable and even uncommonly marked variations in the frequency and regularity of the heart's action, the changeable quality of the pulse, the frequency with which respiration, likewise, assumes an intermittent or arrhythmic type, show that there is no equilibrium between the exciting and inhibitory powers of the nerve centre during the first week of life. The defective inhibitory power of the vagus has been experimentally demonstrated in animals, by Soltmann. That the inciting and regulating influences of the brain, and more particularly of the cerebrum, should still be imperfect at birth, is readily conceivable, if it be borne in mind that considerable areas of the brain receive their myelin only at a later and very variable period of infancy. Then, while the function of nerve fibres may not be entirely dependent upon the presence of the myelin element, its absence must nevertheless limit their conducting power to a considerable extent, and seriously compromise the physiologic accomplishments of all nervous function. We know, not alone from the actual evidence which pathology affords, but also from the results of animal experimentation and neurologic examination of the newborn, that destruction of myelin is followed by disturbances of greater or lesser intensity. In animals (cats, dogs, etc.) blind at birth, the closure of one eye delays the appearance of myelin in the corresponding optic nerve (Held). The optic nerve of a child born at the eighth month, shows, a month later, a much greater proportion of myelin than that of a child born at term (Flechsig). The spinal cord, the cerebral centres and nerve tracts which govern the first (reflex) external manifestations (fœtal movements) of life, and in the newborn, vegetative growth, the acts of sucking and tasting, and soon, the instinctive recognition of food, etc., are the first to receive their myelin constituent, as was previously stated. The fact that at birth, extensive areas of the brain are devoid of myelin, certainly affords a plausible explanation of the absence, or at least inadequacy, of its function.

The brain of newborn animals reacts differently to electrical stimulation from that of the fully grown. The irritation of both motor and sensory centres produces an entirely different effect (Steiner).

Then, in the greater necessity for sleep (about 20 hours a day during the first weeks, 13–15 hours by the end of the first year), we have a good criterion of the peculiar structural character and functional inadequacy of the child's central nervous organs. From the standpoint of hygiene of the nervous system in infancy and early childhood, it cannot be too often repeated, that during the first months of life, not only every strong and shrill or glaring (acoustic, optic, etc,) impression, but also any irritation of milder degree, if uniform in intensity and long continued, wears out, indeed exhausts, the brain. Again, when as a consequence of certain, and by no means infrequent, irregularities in development, special aptitudes become manifest at an unusually early age, is it most undesirable to further develop and cultivate them; almost invariably does such effort prove detrimental to the general condition of the nervous system. An interesting insight into the functional inadequacy and peculiarily of the child's nervous system may also be obtained by testing the *reflexes.* The tendon-reflexes, the knee jerks more particularly, are already appreciable in the premature infant; from the second month to the second year they are distinctly livelier than in healthy adults. With the exception of the first week of extra-uterine life, when it is more often absent, the abdominal cutaneous reflex is likewise most active in infancy; the same may be said of the plantar reflex. A striking peculiarily in this connection, is the presence, until from the sixth to the tenth month, of the Babinski reflex; a phenomenon of pathologic significance in the adult, but which, at this period of imperfect cerebral function, represents the normal reaction. Oppenheim's reflex (Unterschenkelphanomen) may likewise be elicited in healthy infants. The reflex closure of the eyelids on the approaching of a finger, occurs already at birth, is particularly active during the first month and often has a clonus-like character. Winking, on the other hand, which is a true optic reflex, does not occur in the newborn and is rarely observed before the sixth to the eighth week. The pupillary reflex to light, which is already appreciable in the prematurely born, shows particular activity and amplitude (more especially in girls) in the latter phase of infancy. A distinct reaction to accommodation the author has only obtained after the fourth week. The very fine oscillations of the pupil (Psychoreflexe), which, as is well known, are almost constantly perceptible and represent the reaction determined in the organism by the constantly varying psychic and sensory impressions (Rieger, von Forster, Laqueur, etc.), become manifest in children (individual precocity occasionally presenting) only after the third month.

Our knowledge of the psychology of the first phase of life is proportionately meagre and uncertain, although much can be derived from the study of the above-described peculiarities (electrical reaction, reflexes, etc.) as regards objective points of difference between the functional activity of the child's nervous system and that of the adult. The individual himself in later years has no recollection of the mental processes which took place in his own brain during the very first years of life; therefore, in making any statement concerning the dawn of the child's moral existence, we must rely solely on conclusions drawn from analogy, which, necessarily, are of doubtful value.

The exact age at which conscious *mental operations* begin, is absolutely unknown to us. Even in intra-uterine life, very likely, vague psychic influences come into play, in connection perhaps with fœtal movements. The more vulgar forms of sensation (preferably hunger, thirst, uneasiness) are already developed, at least in a rudimentary fashion, from the very earliest moments of life and constitute the underlying causes of various instinctive reflex manifestations (unyielding movements, crying, sucking). That these vital manifestations of the newborn (sucking, swallowing, ocular movements), which to us might appear to depend upon volitional activity, are nothing more than inherited reflexes, instinctive performances, is indeed hardly questionable, if we take into consideration the primitive state of the central nervous organs. Tactile impression, perceptions of smell and taste are the first (in the very first days following birth) to graft themselves upon the child's brain, to form there certain connections and association, the first, finally, to ensure recognition (more particularly of food) and to even determine the early appearance of useful movements of defense, as illustrated in Preyer's observation.

The aversion to light which is so manifest at birth, disappears only from the tenth to the twentieth day. The incoördinate, atypical movements of the eyeballs which turn towards light under purely reflex influences, become coördinate and regular after a few weeks. It is only after the fourth or fifth week however, that the child distinctly fixes the eyes upon objects placed before it, and scarcely before the third or fourth month, can it be trained to notice an object presented at the periphery of the visual field, and to further control the degree of perception by following it with the eyes.

The newborn is deaf; acoustic stimuli call forth a response only coincidently with the gradual appearance of other reactions (turning of the head, looking towards), but more and more does the child learn, with regard to hearing as with seeing and feeling, to store up distinct memories of this sense, and by the end of the third month, it has acquired almost perfect control over the use of all its sensory aptitudes. In the course of the following months, during which the child's mental disposition (its likes and dislikes) becomes ever more sharply defined, and the faculty of observation and of associating impressions progressively develops, consciousness of self-materialness gradually asserts itself; the outer world stands out more and more prominently as something essentially distinct from the *ego*. At the end of the first year, speech begins to develop (with varying rapidity in different individuals), and from that time on, the child enriches its knowledge and deepens its judgment, under the influence however, of its surroundings. The effect of new impressions in varying previously acquired notions, is dependent in great part upon the child's psychic inclinations; advance is further promoted by a pronounced distaste for unclear situations (a fact well illustrated by the tendency in children, especially between the ages of five and ten, to ask innumerable questions).

If, however, the psychic conformation thus acquired is essentially as adequate as the psychic mechanism of the adult, the child's mind nevertheless differs from that of the latter, for a considerable length of time, by the greater mobility of the feelings, the greater influence of the latter upon both the sequence and course of ideas, finally, by the lack of uniformity and purpose displayed in regulating and grouping efforts, whether of simple demonstration or of resistance. Owing to ever increasing new perceptions, the sexual impulses, which appear (not infrequently as early as the tenth or eleventh year) as rather unclear perceptions at first become ever more clearly defined later. The child's moral existence suffers, at the time of puberty, a shock of greater or lesser intensity, a jar, with the gradual adjustment of which (determination of character), the childlike or youthful mind first acquires its proper physiognomy and then gradually transforms itself into that of the mature man.

ORGANIC DISEASES OF THE NERVOUS SYSTEM*

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SECTION I.

CONGENITAL DISEASES OF THE NERVOUS SYSTEM

1. *Acephalia, Amyelia.*—The greatest disturbance of the central nervous system consists in the total absence of the brain and spinal cord. The vertebral column remains open posteriorly, the base of the skull is imperfectly developed, and there is complete absence of the calvarium. In amyelia the spinal ganglia and sensory roots are present and the latter exhibit an attempt to join the imperfectly developed spinal marrow; muscles are also present, as well as those portions of the peripheral nerves which are derived from the spinal ganglia. Monsters of this type are not viable and die in the fœtal period (cases of Manz, Leonowa, Petrén, Gade, etc.). Cases have also been observed with partially preserved spinal marrow (Wolfram).

2. *Anencephalia (Hemicephalia).* The term anencephalia [hemicephalia is not a good term on account of the analogous use of the prefix for unilateral disturbances of the cerebrum (Sternberg)] is used to describe a malformation in which the spinal marrow, the medulla oblongata and portions of the basal ganglia are present, while the cerebral hemispheres are absent and the cerebellum is usually atrophic. There is no calvarium and the cranium is closed by a spongy, vascular mass which sometimes exhibits nodular swellings and prominences simulating cerebral convolutions (area cerebrovasculosa, Recklinghausen). Microscopic examination of such cases has shown that the parts of the central nervous system which are present are, in the main, well developed, although individual systems of fibres are absent or imperfectly formed (pyramidal tract, lateral cerebellar tract); while, on the other hand, certain nerve tracts, when the parts which they are intended to supply are absent, may be abnormally developed and run in abnormal directions. The neurologic significance of these microscopic findings was recognized and elaborated by v. Monakow and his students. The peripheral nerve organs are developed even when their connections with the central organs are absent (autodifferentiation, Roux).

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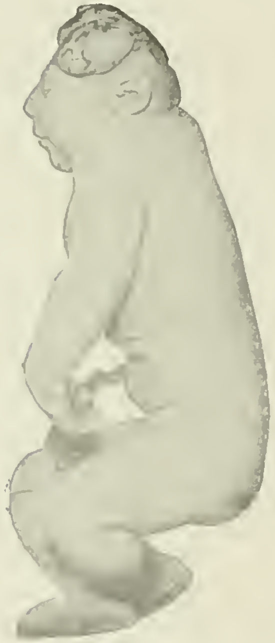
* Except diseases of the meninges.

It is possible that anencephalia is produced by cleft-formation in early fœtal life or by failure of the medullary tube to close; possibly the malformation is due to certain chemical abnormalities within the ovum. At all events, it has been possible by introducing chemical or toxic substances into the ova of animals to prevent closure of the brain-plate (Hertwig, Roux, Feré). Other forms of malformation of the central nervous system have been produced in animals by mechanical means (Dareste, Tichomirow, Kollmann). Several cases of anencephalia have been observed in the same family. The presence of anencephalia does not necessarily involve death of the fœtus. It may be born at the normal period and survive for several days. Sternberg and Latzko in their physiologic studies of an anencephalic monster which lived three days found that crying, sucking, defecation and urination, the corneal reflex, sensation of pain and discomfort, mimic reflexes, movements of the extremities and tendon reflexes in the arms and legs were present. Evidently these vital manifestations may yield valuable information in regard to the seat of reflex processes in the nervous system.

Fig. 23.



Anencephalia. Still-born child.

Anencephalia is often associated with other malformations affecting not only the spinal marrow (alterations in the central canal, etc.), but other organs also (aplasia of the adrenal bodies). There appears to be a preponderance of females among anencephalic monsters.

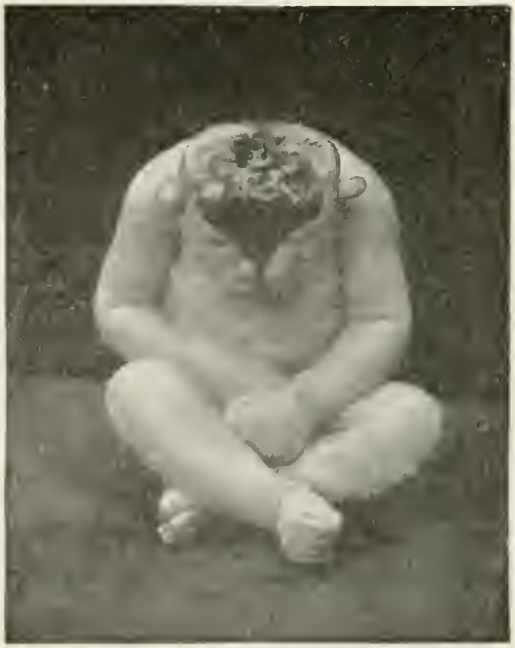
3. *Cyclopia* (*Cyclencephalia*).—In this malformation the brain canal is closed but the cerebrum exhibits marked alteration. The cerebral hemispheres are poorly developed and not separated from one another. The absence of symmetry in the nervous organs shows itself most markedly in the presence of a single eye in the median line, which is usually very poorly developed. The cerebrum shows either cystic degeneration (Nägeli) or arrested development (Leonowa). The optic thalami and portions of the base of the brain sometimes exhibits a reduplication of the usual or normal embryonic structure. With cyclopia are occasionally combined malformations of the skeleton of the face (arrhincephalia, Kundrat), in which the nose may exhibit a variety of bizarre, snout-like deformities; children with this malformation are not viable (cases of Kundrat, Nägeli, Monakow, Falk, Leonowa, etc.).

Congenital fusion of the two cerebral hemispheres without other malformations has also been described; the abnormality does not interfere with the infant's bodily development (Seeligmann).

4. *Porencephalia.*—This term indicates a loss of substance in the cerebrum (Heschl, Kundrat), causing funnel-shaped retraction of the brain and sometimes leading to the formation of cysts. The abnormality is most frequent in the region of the fissure of Sylvius, corresponding to the distribution of the arteria fossæ Sylvii. Not infrequently (in 30 per cent. of the cases according to Siegmund) both hemispheres are affected. The porencephalous hemisphere is usually retarded in its development and weighs less than its fellow. (In Acker's case the sound hemisphere weighed 670 and the diseased hemisphere 411 grams.)

Porencephalia is a purely anatomic condition and may be congenital or acquired. In the latter case it represents the result of an acute cerebral process, an encephalitis, a meningo-encephalitis, a hæmorrhage, or embolic softening. Opinions are divided on the question whether congenital porencephalia represents the remains of an intra-uterine morbid process, as formerly believed by Kundrat, or whether it is a developmental disturbance, *i. e.*, a malformation in the narrower sense of the term (v. Kahlden). According to the first view, portions of the cerebrum are deprived of their nutrition early in the fœtal period as a result of inflammation or disease of the vessels (embolism), causing anæmic necrosis, degeneration or absorption of the affected portions of the cerebrum. The arguments in favor of a primary developmental anomaly are the seat of the anomaly, which is fairly constant; the regular arrangement of the convolutions in the region of the defect; the microgyria; and especially the association with other malformation of the central nervous system, particularly unilateral deformities of the skull. Certain very careful investigations of Zingerle go to show that at least a certain proportion of the cases of porencephalia are due to intra-uterine cerebral disease.

Fig. 24.



Anencephalia. The illustration shows cleft-formation in the face. View from above, showing the area cerebro-vasculosa quite distinctly.

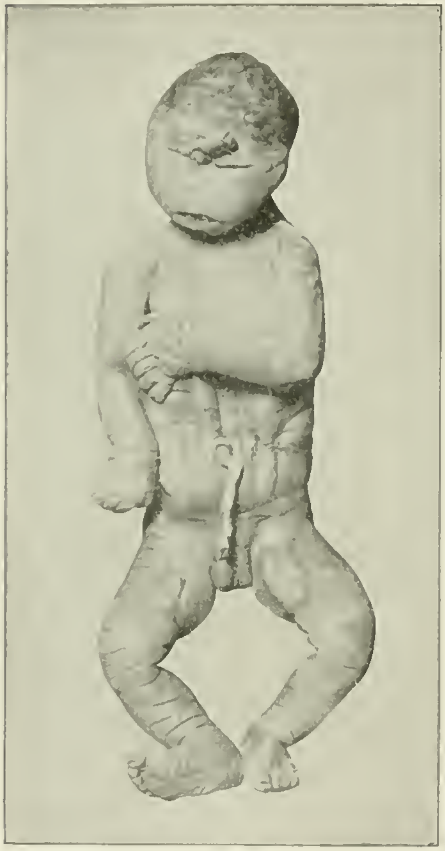
Clinically, porencephalia usually corresponds to a spastic hemiplegia or diplegia associated with idiocy, epilepsy and disturbance of speech.

The presence of these symptoms does not, however, justify the diagnosis of a porencephalous defect in the cerebrum. The clinical significance of porencephalia will be considered again in connection with the cerebral palsies of children.

5. *Microcephaly.*—The term microcephaly embraces a number of congenital diseases of the cerebrum in which the brain constantly, and the skull sometimes, are smaller than normal.

The many cerebral malformations, representing a great variety of different forms, which are included under this name have been carefully sifted by Giacomini and divided into pure microcephaly and pseudomicrocephaly. The former represent a general malformation or arrested development of the cerebrum; the latter, intra-uterine disturbances affecting the growth of the cerebrum, the result of severe diseases. Searching anatomical investigation of microcephalous brains has shown that the cases belonging to the first group are much more rare than was at first believed. In these cases the brain is not a miniature edition of a normal human brain, since other abnormal formations, such as macrogyria, arrested development of the forebrain in comparison with that of the midbrain and hindbrain, and animal types of convolutions are present. Most of the cases regarded as microcephaly exhibit on section sclerotic and degenerative processes, cysts and hydrocephalus, so that the changes in the cerebrum must be regarded as the remains of intra-uterine diseases. As a rule, the weight of these microcephalous brains is considerably below the normal (see statistics collected by Pfleger and Pilcz).

Fig. 25.



Arrhincephaly with absence of the eyes (belonging to cyclopia). From the collection of photographs in the gynecologic clinic of Dr. Hofrat Schauta in Vienna.

In microcephaly changes in the calvarium are the rule, making the diagnosis of microcephaly clinically possible. The alteration consists in a reduction of the circumference of the skull and in a change of shape, the skull being usually spherical and markedly brachycephalous; the forehead is flat and receding, and the occipital portion is feebly developed. As the skeleton of the face is well formed and prominent, a bizarre appearance (bird-face) may result. The measurements of the microcephalous skull are shown in the following table:

Circumference of the Skull.

In the normal child (after Bendix)		In the cases of microcephaly (Pfleger and Pilcz)	
End of the 1st month	35.4 cm.	14th day	22.0 cm.
End of the 3rd month	40.9 cm.		
End of the 6th month	42.7 cm.		
End of the 9th month	45.3 cm.	38th week	28.0 cm.
End of the 12th month	45.6 cm.		
End of the 15th month	46.2 cm.	15th month	30.5 cm.
End of the 18th month	46.9 cm.		
End of the 21st month	46.8 cm.		
End of the 2d year	48.0 cm.	2d year	40.0 cm.
End of the 3rd year	48.5 cm.		
End of the 4th year	50.0 cm.	4th year	40.0 cm.
End of the 5th year	50.0 cm.	4th year	43.0 cm.
End of the 6th year	50.9 cm.	4th year	44.0 cm.
End of the 7th year	51.0 cm.	4th year	44.0 cm.
End of the 8th year	51.3 cm.	6th year	44.0 cm.
End of the 9th year	51.7 cm.	7th year	47.0 cm.
End of the 10th year	51.8 cm.	9th year	44.0 cm.

Microcephaly however occurs also in association with a skull of normal circumference.

The question of the ossification of the calvarium is of practical importance. While it was formerly believed that the interference with the growth of the cerebrum consisted primarily in the premature ossification of the skull (probably because of Virchow's erroneous belief that the sutures close prematurely in cretinism); it is now well known that the primary lesion is situated in the brain itself and that failure of the fontanelle to close, abnormal persistence of the frontal suture, deficient ossification of the calvarium and even persistent separation of the sutures occurs later in life in microcephaly.

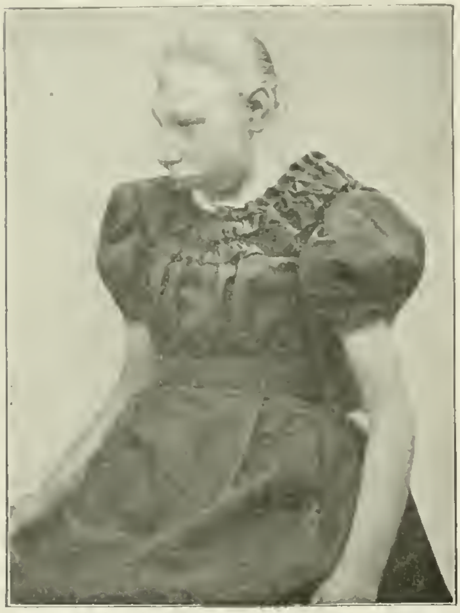
Microcephaly does not interfere with the individual's bodily development. The subjects may attain the age of fifty years and over. On the other hand, microcephaly is usually accompanied by a high degree of idiocy. Many of the children who even in infancy arouse attention by their restlessness, their inability to concentrate, total absence of any power of reasoning, absence of reaction to pain (Thiemich), are microcephalic. Sometimes there is general muscular flaccidity and the child cannot raise its head. Much more frequently marked muscular rigidity is the most prominent symptom. The arms are tightly pressed against the chest, the elbows bent, the hands are flexed on the forearm and the fingers turned into the palms; the legs are in extension and extreme adduction, with a tendency to crossing. The trunk muscles may be so rigid that the child can be picked up like a piece of wood.

The abdomen is often of board-like hardness and retracted or scaphoid. Sometimes this picture of microcephalic rigidity with idiocy (Freud, Ibrahim) is complicated by athetoid movements, dysphagia, pseudobulbar symptoms and epileptic attacks.

Ibrahim attempted to distinguish clinically between pure microcephaly dependent upon arrest of development and pseudomicrocephaly due to some intra-uterine disease. Although this distinction cannot be rigidly maintained, it appears nevertheless that athetosis, bulbar symptoms and epilepsy point rather to pseudomicrocephaly while general rigidity indicates a pure microcephaly.

The **prognosis** of microcephaly is extremely gloomy. Functions which are often preserved in children with irreparable injuries to the cerebrum, are usually completely lost in microcephaly. It is practically impossible to effect any improvement in the child's mental state, to accustom it to cleanliness or even to get it to use its legs.

Fig. 26.



Microcephaly. Half grown girl with pronounced flattening of the occiput and bird-face.

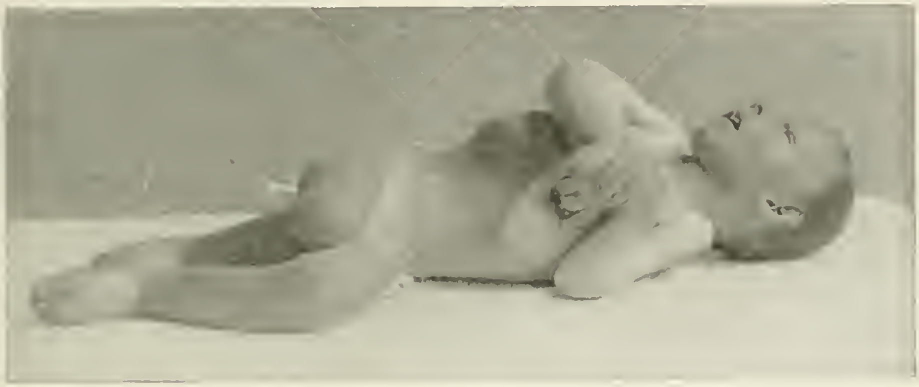
For a time the therapeutic outlook seemed more hopeful when Lannelongue, starting out with the idea of a primary ossification of the skull, attempted by chiseling out large pieces of the calvarium to give the cerebrum an opportunity to develop. Operations of this kind have since been repeatedly performed, and whole sections have been removed from the circumference of the skull, so that the calvarium was connected with the lower portions of the skull only by the soft parts; but, although the children stood the operation surprisingly well, it was not followed by the desired result (Löwenstein) and the operation must now be regarded as obsolete (Pilcz).

6. *Congenital Hydrocephalus.*—The term hydrocephalus is used in pediatric neurology to designate a group of morbid conditions having as a common symptom enlargement of the skull, the result of accumulation of fluid. The term is used with little regard to the fact that the pathology and etiology of hydrocephalus may be exceedingly variable; the one distinction made being between external hydrocephalus (effusion between the surfaces of the brain and the calvarium) and in internal hydrocephalus (the collection of fluid in the ventricles of the brain).

Clinically the cases are divided between acute and chronic hydrocephalus. It would be well to give up this comprehensive term, which, like "cerebral infantile palsy," merely tends to the making of symptomatic, instead of etiologic diagnoses, in favor of a rational classification based on the causes of hydrocephalus.

It cannot be denied that such a classification would be difficult in the present state of our knowledge. The following, which is in part copied from Leon d'Astros, may be suggested: (1) *Congenital hydrocephalus.* This is usually internal, rarely external, and belongs to the group of congenital diseases of the central nervous system now under discussion. (2) *Acquired hydrocephalus.* This may be acute or chronic. The acute form belongs among the inflammatory diseases of the meninges. Chronic hydrocephalus may also be the expression or result of diseases of the external and internal membranes of the brain. Among these simple meningitis and ependymitis due to hereditary syphilis are the most important. Chronic acquired hydrocephalus is also observed after sinus phlebitis (Marfan), in sclerotic processes affecting the brain, in brain tumor, and after severe diseases generally. In all these cases the fluid may accumulate within the ventricles or on the outside of the brain surface.

FIG. 27.



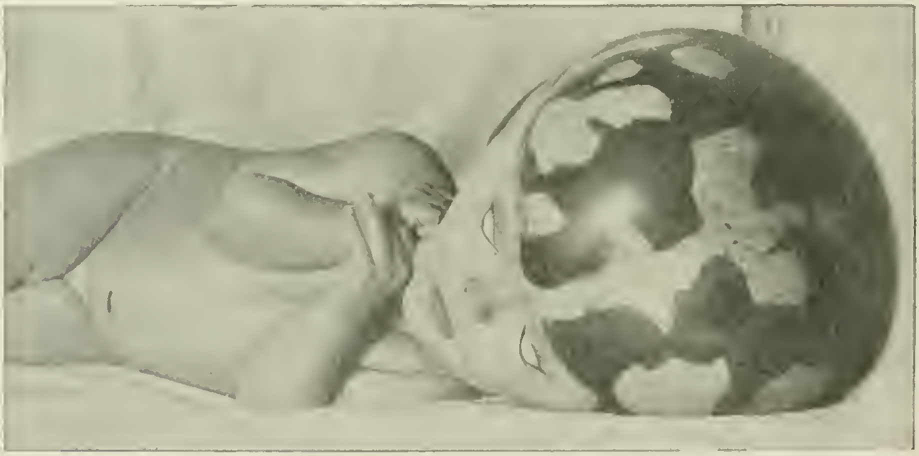
Microcephaly with idiocy. Boy four years old. Unilateral convulsions; scaphoid abdomen; circumference of head 41 cm.

As the various forms of hydrocephalus will be discussed in this work in their appropriate places, we shall confine ourselves here to a description of congenital hydrocephalus.

Congenital hydrocephalus in the great majority of cases is *internal;* although, in view of the statements of Leon d'Astros, Heubner and Bókay, the occurrence of congenital *external* hydrocephalus which had been previously denied, can no longer be called in question. In the latter form the brain is intact, usually somewhat diminished in size, and also contains an accumulation of fluid within the ventricles; the condition probably represents the remains of an intra-uterine meningitis. External hydrocephalus also occurs after intra-uterine shrinking processes and in combination with defects and malformations of the brain.

Congenital *internal hydrocephalus* also suggests the previous existence of intra-uterine disease affecting the vascular apparatus, with excessive exudation of fluid—either a hypersecretion of fluid from the chorioid plexus due to unknown causes or actual inflammatory processes. If this collection of fluid takes place at an early stage, the development of the brain is interfered with (see below; hydromicrocephalus); in that case hydrocephalus has the same significance as arrested development of the brain. In other cases the accumulation of fluid brings about atrophy of the already completely developed brain.

Fig. 28.



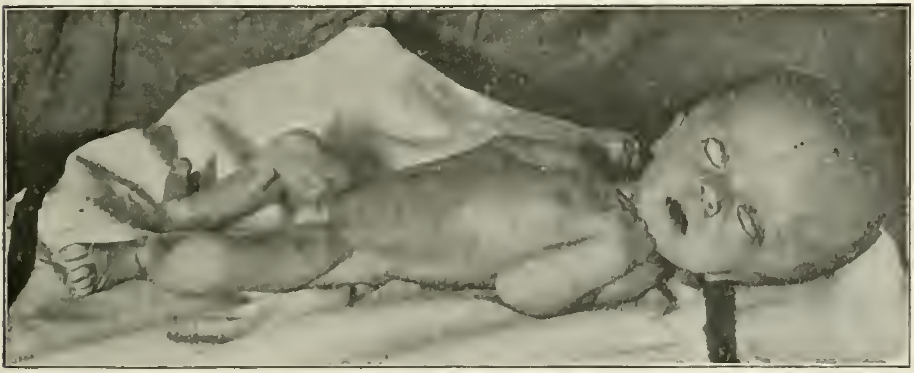
Internal hydrocephalus of enormous extent. Circumference of head 75 cm. Child of eight months. The shaded portions of the skull represent the remains of the bones. The eyes are dislocated downward.

The causes of congenital hydrocephalus are by no means certain. Alcoholism, tuberculosis and nervous diseases on the part of the parents have been assigned. The influence of syphilis, which undeniably plays a part in acquired hydrocephalus, is doubted in the case of the congenital variety. Possibly the condition represents a parasyphilitic affection (see below). Hereditary hydrocephalus has been described and the disease may occur in several members of the same family.

The *quantity* of fluid in hydrocephalus may be enormous (5 litres and over; usually the quantity is about one litre). The fluid is watery and contains no cellular elements, and chemically corresponds to an indifferent saline solution; the low percentage of albumin and the large quantity of salt in the fluid are characteristic features. The greatly distended ventricles exert pressure on the mantle of the brain, causing flattening of the convolutions, narrowing of the soft cerebral mass and, in severe cases, complete obliteration, leaving nothing but a thin layer which can be recognized only with the microscope. The basal ganglia are often flattened; the cerebellum is altered and displaced (Chiari).

As ossification does not take place, the soft skull is unable to resist the pressure of the fluid within the brain, it becomes enormously enlarged, the bones of the skull are attenuated, the sutures greatly widened, and the fontanelles bulge. The circumference of the head has been known to reach 50 and even 100 centimetres. In these cases the bones of the skull are widely separated and appear like islands in the connective tissue, which spans the fontanelles and sutures. The cranial veins are usually dilated, the skull is smooth and scantily covered with hair. In contrast to the enormously enlarged skull, which projects on both sides of the head, the countenance appears diminutive, like a mere appendage. The eyes are usually dislocated downwards, bringing the upper border of the cornea into view. The expression is staring and devoid of intelligence and the movements of the eyes are usually restricted. The abnormal position of the eyes is probably due to the pressure of the fluid on the attenuated, unresisting orbital plates. Nystagmus, strabismus, sluggishness of the pupils and atrophy of the optic nerve are sometimes present.

Fig. 29.

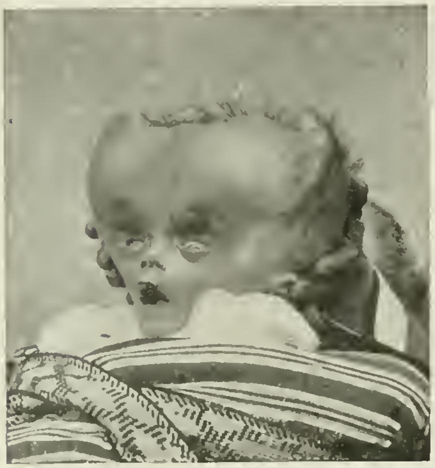


Internal hydrocephalus. The downward direction of the eyes and the contractures in the arms are well shown.

The mentality of these children ranges between extreme idiocy and normal intelligence. Most of the children are feeble-minded, apathetic, and have more or less difficulty in speaking. Motor disturbances are usually well marked and manifest themselves in spasms, paresis, universal rigidity, tremors, awkward movements of the hands, etc. As the lower extremities are almost always more severely affected than the upper, walking is very much delayed; there is a tendency to crossing of the legs and the gait is spastic. Sometimes, the spasms and paresis are more marked on one side than on the other. The reflexes are increased. Aside from these motor disturbances, hydrocephalic children exhibit a peculiar indolence, a disinclination to exercise, which is enhanced by the difficulty of steadying the abnormally large and heavy head. Twitching of individual groups of muscles and general convulsions are by no means rare. Hydrocephalic children usually present a distinctly delicate appearance; they are pale, emaciated, with senile expression, cry a good deal and do not increase properly in weight. The appetite is not bad, but digestion is usually retarded.

The circumference of the skull may continue to increase for some length of time after birth. Sometimes the enlargement takes place in successive stages, each increment being accompanied by brain symptoms. Spontaneous evacuation of the cerebral fluid through the nose, has rarely, if ever, been observed in children.

Fig. 30.



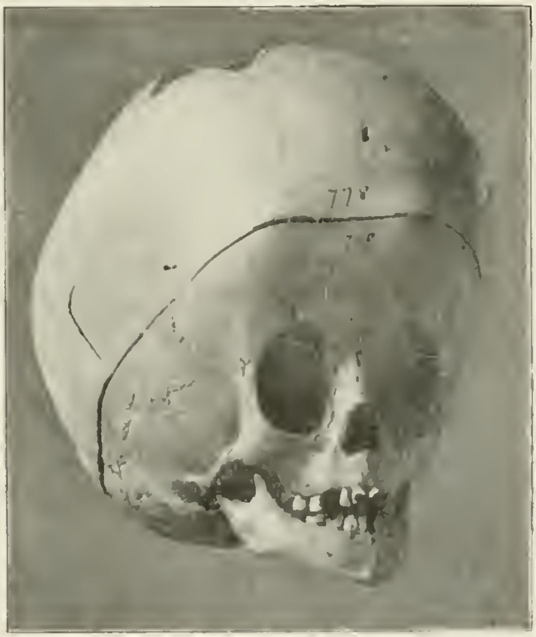
Hydrocephalus with spontaneous rupture. The hydrocephalus developed after accidental healing of a congenital spina bifida. Greatest horizontal circumference 54 cm. Clear serous fluid to the amount of about 3 litres was evacuated. The child was eight months old and lived 8 days after the rupture had taken place. The photograph was taken while it was yet alive. Observation by Dr. Veninger-Meran.

The **prognosis** of congenital hydrocephalus depends primarily on the severity of the condition. In milder cases, in which the clinical symptoms are not too pronounced, there is more hope of recovery and especially of intellectual development than in a case of excessive hydrocephalus in which walking is greatly delayed and the mental functions are permanently impaired. A large proportion of these children die soon after birth or in early childhood.

The **treatment** of hydrocephalus is not encouraging. In view of the favorable results obtained with iodides and mercury in many varieties of acquired hydrocephalus, these remedies should be tried in congenital cases. Various operations have been devised for the purpose of diminishing the quantity of fluid or preventing its re-accumulation. Different methods of puncturing the ventricles, followed by the injection of iodine or the insufflation of sterilized air, the application of permanent external drainage and, finally, the establishment of a permanent communication between the ventricles of the brain and the meningeal space or the external integument have been tried in the hope of accomplishing the above result. The pressure bandages which at one time were extensively employed have now been practically abandoned and are used only as part of the after-treatment following evacuation of the fluid. There can be no doubt that cases of hydrocephalus have been improved by repeated *lumbar puncture,* but permanent results are extremely rare and the effect of the procedure in most cases is only temporary. Acute cerebral manifestations with the symptoms of a recent meningitis are not rare after evacuation of large quantities of fluid and are probably due to speedy re-accumulation of serum. Nevertheless, especially in cases of increasing pressure, as, for example, when inflammation of the optic nerve goes on to atrophy, lumbar puncture should be tried; but as in some cases the communication between the ventricles of the brain and the spinal canal is closed, the operator should not be surprised if he fails to obtain a considerable flow of cerebrospinal fluid.

Hydromicrocephalus or hydranencephalus (Cruveilhier) is the term used to designate a rare malformation in which the external skull is diminished in size instead of enlarged, and the brain itself is represented by a watery cyst. The condition is apparently due to the collection of hydrocephalic fluid at a time when the cerebral cortex is still in the initial stages of its development. The medulla oblongata and small portions of the basal ganglia may in such cases represent the sole remains of the brain. In spite of these marked alterations clinical symptoms may be wanting. In a case described by Zappert and Hitschmann, although the child lived 11 days, deficiency of the brain had not been suspected.

FIG. 31.



Hydrocephalic skull. Striking disproportion between the bones of the skull and those of the face.

7. *Hypertrophy of the brain* is an alteration of the brain manifesting itself by excessive weight and increase in bulk. The convolutions are flattened, and the ventricles are usually totally obliterated (Schick) or they may occasionally be dilated (Anton). The microscopic findings are normal; occasional persistence of the thymus gland in adult individuals and aplasia of the adrenal bodies are notable features. The skull is sometimes, but not always, enlarged; the shape, however, is not that of hydrocephalus. The bones of the skull are thin and porous.

This hypertrophy of the brain is at the present time regarded by most authorities as a congenital condition, it being doubtful whether it can occur as the result of external injuries (lead poisonings, Barthez and Rilliet). It is possible that heredity is a feature in the etiology.

The condition does not necessarily cause symptoms. Indeed, it is difficult to draw the line between normal and pathological size of the brain, for we know that many persons of great mental powers have extremely heavy brains. Quite often the existence of a cerebral anomaly is only discovered at autopsy. When symptoms are present they are due to the pressure of the skull on the surface of the brain. The most important are convulsions, which may go on to the status epilepticus and are brought on through reflex irritation (auditory and visual impressions). Intelligence is sometimes diminished; in severe cases deep coma, with protracted convulsions, and death occur. There are no local brain symptoms. It is worthy of note that the morbid symptoms as a rule do not develop until after the first year of life, because up to that time the skullcap is soft enough to yield to the internal pressure. From hydrocephalus, which may present very similar symptoms, the condition according to Schick can usually be differentiated by the negative result of lumbar puncture (due to the absence or diminution of the cerebrospinal fluid).

Rachitic subjects occasionally present at the autopsy, brains of abnormal size and density when there have been no clinical symptoms during life. Whether this hypertrophy of the brain is to be regarded as identical with the above-described form is not decided. In fact, the clinical as well as the pathological picture of hypertrophy of the brain is so imperfectly known that there is very rarely any question of making a differential diagnosis at the bedside.

8. *Defects in the skull and the vertebral column with protrusion of the nerve substance* (*Cerebral and spinal herniæ*). It has already been emphasized that the highest degrees of separation, or rather imperfect closure of the braincap and of the medullary tube, produce marked malformations of the central nervous system. Such cases are to be regarded as curiosities. More practical interest attaches, however, to those cases in which separation and failure to close of the central nervous system and its bony envelope are confined to circumscribed areas. Failure of the skull to ossify may manifest itself in the substitution of connective tissue membranes for bony portions of the skull (cranial defects, Heubner, Engstler); indeed, ossification may be entirely absent and there may be merely a membranous skull (Stilling). As a rule, however, failure of the bony envelope to close is associated with marked changes in the central nervous system itself and protrusion of individual portions of the brain and cord through the abnormal openings. We then have to deal with herniæ of the brain and spinal cord.

(*a*) Hernia of the brain (*Cephalocele*).—Protrusion of the brain substance may take place through an artificial opening in the skull (trephining, injuries) or through defects due to disease (meningocele spuria, Billroth). The hernia represents a pulsating, cystic, compressible tumor and usually requires surgical treatment (Bayerthal).

A more important condition from the pediatric standpoint is congenital hernia of the brain.

Intermediate between the above-mentioned forms of acrania and anencephalus, which are to be regarded as the most extreme examples of separation of the skull, and hernia of the brain, is *exencephalus*, a condition in which a variable portion of the brain escapes through a large opening in the misshapen skull (Muscatello). Other developmental disturbances of the brain are also present and the child is not viable (Lyssenkow).

The difference between true hernia of the brain (cephalocele) and exancephalus is that in the former condition the defect in the skull is circumscribed. By far the greater portion of the brain is normal, and the cerebral functions as well as the viability of the infant do not appear to be materially impaired. The contents of the hernial sac consists either of cerebral substance with a cystic and dilated portion of the ventricle (encephalocystocele) or a meningeal protrusion containing fluid (meningocele).

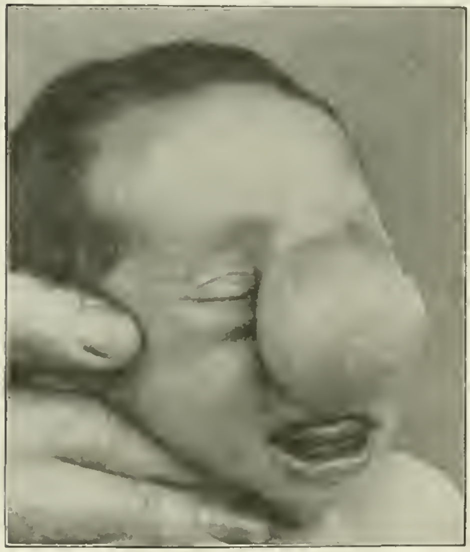
Of the two, *encephalocystocele* is the more frequent and more important form of brain hernia. Its most frequent seat is the nape of the neck (cephalocele occipitalis) and the hernia contains altered cerebellar substance and the distorted fourth ventricle. Another seat is the root of the nose (c. nasofrontalis, c. naso-orbitalis) or the sagittal suture (c. sagittalis); in these cases the contents consist of cerebral substance, and the cavity of the cyst communicates with one of the ventricles. The tumors are usually, but not always situated in the median line. The dura becomes attached to the periosteum at the edge of the bone defect. The cerebral substance, as in hydrocephalus, exhibits every degree of contraction down to a mere membrane covered with nerve cells. The vessels are numerous and dilated. The external coverings either consist of normal skin or represent attenuated glistening membranes, sometimes exhibiting scars from intra-uterine processes of repair. The tumors range in size from that of a walnut to that of a child's head. The consistency is soft, elastic and usually fluctuating; sometimes the tumor is transparent; the structure is often lobulated and harder portions can be felt here and there. When the child cries or the tumor is compressed, pulsation and distention are often, although not always, present. The defect in the bone can often be felt.

Clinical symptoms are not necessarily present in every case and are particularly apt to be wanting when the hernia occupies the anterior or the upper portion of the skull. The most important symptoms are idiocy, nystagmus, strabismus and atrophy of the optic nerve. Other malformations, particularly in the central nervous system, are frequently present (hydromyelia, spina bifida, etc.).

Meningocele is a rarer condition and is difficult to distinguish from the above-mentioned variety. The defect in the bone is usually smaller. Nervous symptoms, particularly atrophy of the optic nerve, are entirely wanting. Horsley suggests the original expedient of determining the presence of reacting cerebral substance by means of irritation with the electric current.

The **prognosis** of hernia of the brain is unfavorable inasmuch as children with large tumors not infrequently die from secondary infection of the tumor. The surgical treatment of these conditions has been greatly perfected during recent times (v. Bergmann) and, in the case of hernias situated in the nasal region, consists in the complete removal of the protruding sac. When the hernia is situated in the occipital region, the sac is opened and the protruding portion of the brain replaced. The opening in the skull may be closed by an osteoplastic operation or with a Fränkel celluloid plate. If the pressure on the brain is severe, lumbar puncture is indicated (Preisich).

Fig. 32.



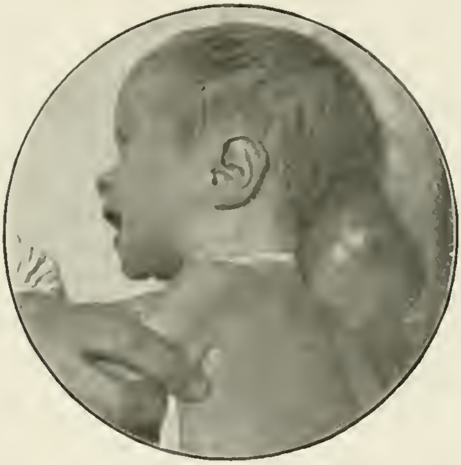
Cephalocele nasofrontalis. Child seventeen days old. At the operation the tumor proved to be an encephalocystocele.

(*b*) Hernia of the spinal cord. (*Spina Bifida.*) Hernia of the spinal cord or spina bifida has far greater clinical importance than hernia of the brain. It represents the most frequent malformation of the central nervous system in children (according to Demme, the incidence is one in 630 births) and has been exhaustively treated in the literature. Our knowledge in regard to these conditions has been greatly extended during the past few years by the important investigations of v. Recklinghausen, which were followed by the valuable contributions of Muscatello, Hildebrandt, Bayer, Bockenheimer, de Ruyter, Wieting, Katzenstein, and many others.

Spina bifida results from failure of the medullary groove to unite at a certain point. This union normally takes place during the early stages of embryonal life. The bony, muscular, fascial and cutaneous coverings, either as a whole or in individual layers, take part in this failure to unite. In order that the contents of the vertebral canal may protrude through this opening there must be a collection of fluid at the corresponding point, hence, the contents of the hernia are always fluid in a typical case of spina bifida. As in the case of hernia of the brain, several varieties of spina bifida are distinguished according to the contents of the tumor: myelocele (or myelomeningocele), myelocystocele and meningocele.

In *myelocele* the medullary tube fails to unite posteriorly, and that portion of the spinal cord which remains uncovered is forced out backward by a ventral effusion. This represents the most severe form of hernia of the cord and, as not only the spinal marrow itself but the spinal meninges, the bone and the external skin remain uncovered, the invaginated mass of spinal marrow is pushed directly through the opening to the surface of the body, and, after undergoing marked changes, forms the dome of the hernial sac. It appears as an oval, dark red, spongy, vascular layer or *zona medullovasculosa*, containing the remains of nerve elements and giving origin to the altered spinal nerves which enter the vertebral canal. Since the spinal marrow somewhere in its course, say in the dorsal portion, undergoes this change, it is evident that both the cerebral and the spinal portions of the spinal marrow must open into the pathologically altered mass and, as a matter of fact, both the afferent and the efferent openings of the central canal can be found in the pole of the hernial sac. Below the *zona medullovasculosa*, toward the base of the tumor, we find a broader, pearly gray, delicate vascular portion resembling the serous coat of the intestine—the *zona epithelioserosa*, which genetically corresponds to the protruded pia. The portion near the base of the tumor consists of skin with dilated vessels and covered with numerous hairs, and is called the *zona dermatica*. This external form of myelocele, which is very important from a developmental standpoint, is rarely encountered in the typical form here described, because the coverings of the spina bifida as the result of intra-uterine maceration and infection occurring soon after birth, are converted into an ulcerated surface, or the hairy portion may proliferate and the entire sac become covered with epidermis. Myelocele is found most frequently in the lumbosacral, more rarely in the

Fig. 33.

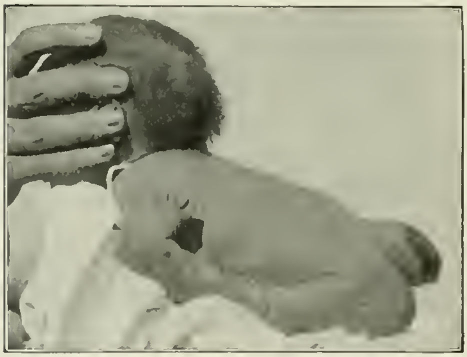


Occipital cephalocele.

cervical region of the spinal marrow. Paralytic symptoms are most marked in this form of spina bifida.

Meningocystocele results from a local dilatation of the central canal, occurring after the spinal marrow is closed, when there is an opening in the bone through which the dorsal portion of the spinal marrow is protruded. In this form of spina bifida the external cutaneous coverings are closed, covering the *area medullavasculosa*, which is adherent to the pia that has also protruded and takes the place of the bulging posterior portion of the spinal marrow. The hernial sac therefore represents the cystic and dilatated central canal and communicates directly with the cavities of the afferent and efferent portions of the spinal marrow. As the ventral portion of the spinal marrow is more or less completely preserved, the motor nerves which take their origin in that portion are normally developed, and palsies of the extremities are less commonly observed in this form of spina bifida. Association with hydrocephalus is frequent.

Fig. 34.



Spina bifida. The demarcation of the zona dermatica is well seen on the spinal sac.

Meningocele is the rarest form of spina bifida. The spinal marrow is closed and there is merely a collection of fluid on the dorsal side of the spinal cord with a saccular protrusion of the pia. The meningocele contains no nerves, or at most a few fibres of the cauda equina may be forced outward by the collection of fluid. Meningocele is usually pedunculated, and it may attain the size of a child's head. The tumor is most frequently situated over the sacrum. Palsies practically do not occur in this form of spina bifida. The frequent association of meningocele with other forms of spina bifida and, more than anything else, the extreme difficulty of recognizing this form of spinal hernia except at the autopsy, renders it doubtful whether all the conditions described under this head are really simple meningoceles. Von Bergmann denies absolutely that meningocele occurs as an isolated affection. In all forms of spina bifida the dura mater is open on its posterior surface and becomes merged in the walls of the hernial sac. Differentiation of the three forms of spina bifida is most important from the standpoint of therapeutics, but unfortunately exceedingly difficult. In quite typical cases the following diagnostic points are of service. *Myelocele* has a broad, sessile base, the outer covering exhibiting the above-mentioned division into three layers; a large opening in the bone is present, through which firmer constituents can be felt. It is practically impossible to replace the tumor, and severe motor and sensory palsies (including paralysis of the rectum and bladder) are observed. *Meningocele* is often pedunculated. The bone cleft is smaller; the tumor is perfectly transparent on lateral trans-illumination, and no solid constituent can be felt. The covering consists of normal skin and the condition is not accompanied by palsies. *Myelocystocele* also has a broad, sessile base; is covered by normal, albeit much attenuated, skin; masses of solid tissue can be felt and, finally, there are sometimes sensory, but rarely motor, disturbances.

In very rare cases hernia of the spinal marrow occurs in the anterior wall of the vertebral column (*spina bifida anterior*).

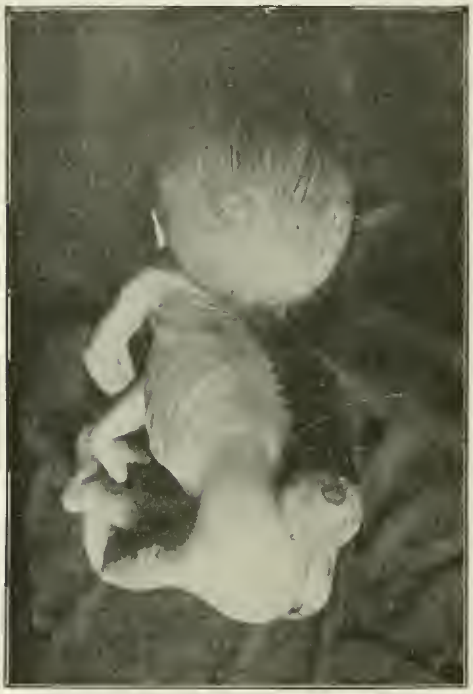
It appears from the above description that the symptoms of spina bifida may be extremely variable. We shall here confine ourselves to those which are observed in myelocele, situated in the lumbosacral region, the most frequent as well as the gravest of the various forms, and concerning which expert opinion is very frequently sought, even in the case of newborn infants. In order to gain a proper understanding of this disturbance we must bear in mind the innervation, which is shown in the following scheme.

I. Lumbar segment, motor	Abdominal muscles, iliopsoas.
II. and III. Lumbar segment, motor	Cremasters, adductors, flexors of the thigh, perhaps sartorius.
III. and IV. Lumbar segment, motor	Extensors, abductors of the thigh.
IV. Lumbar segment, motor	Tibialis anticus, patellar reflex.
V. Lumbar segment, motor	Gluteal muscles, flexors of the knee.
I. Sacral segment, motor	Peroneal muscles, extensors of the foot and toes.
I. and II. Sacral segment, motor	Muscles of the calf, small muscles of the toes, Achilles tendon reflex.
III. and IV. Sacral segment, motor	Perineal muscles (levator ani), muscles of the bladder and rectum.

With regard to the sensory innervation, it will suffice to say that the skin of the greater portion of the upper and inner side of the leg is supplied by the second, third and fourth lumbar nerves; that of the foot and lower side of the leg by the fifth lumbar and second sacral nerves; that of the popliteal space and the posterior aspect of the thigh by the second sacral nerve; that of the perineum, the anus, the mucous membrane of the bladder, the buttocks and the inner side of the thigh by the third and fourth sacral nerves.

It appears from the above that when the spina bifida is situated in the lower lumbar or in the sacral portion of the spinal cord, complete motor (except the iliopsoas) and sensory paralysis of the legs, rectum, bladder and perineal muscles must be present. In addition to the complete atonic paralysis of the legs, this form of palsy is distinguished by obliteration of the anal fold, or even by a funnel-shaped protrusion of the anal region (paralysis of the levator ani and sphincter ani). At the same time the thighs are flexed at the hip-joint at a right, or even an acute angle because the iliopsoas (see scheme) is usually intact. Direct prolapse of the rectum and uterus has also been occasionally observed. Owing to the complete loss of sensation in the skin, so that the infants do not cry when they soil themselves, and probably also on account of trophic changes in the skin deep ulcers often develop in the skin of the buttocks, in the genital region and on the inner aspect of the thigh. All reflexes are completely absent in the legs, although the electric irritability of the muscles, strangely enough, may be preserved. Club-foot is usually present.

Fig. 35.



Spina bifida. Hydrocephalus. Child three months old. The illustration shows obliteration of the anal fold and a suggestion of prolapse of the rectum.

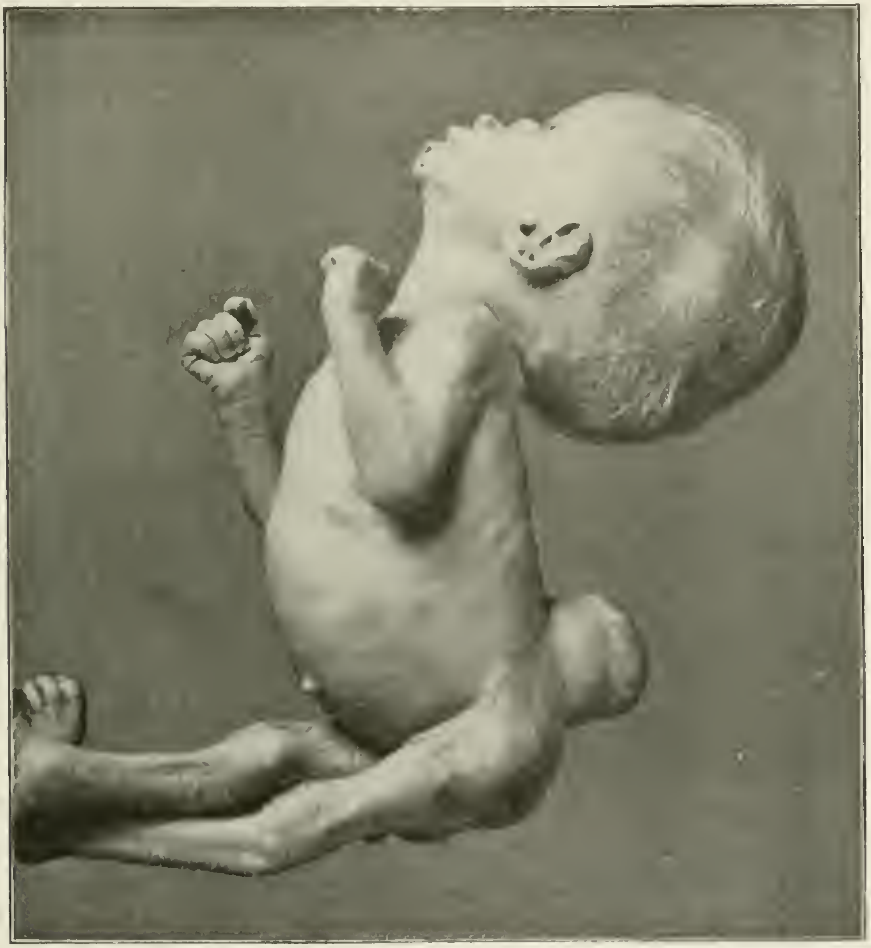
In less severe cases the disturbance is confined to palsies of the foot and of the sphincters.

The combination of spina bifida with other malformations is by no means rare. Thus we find reduplication of the spinal marrow, hydromyelia, dilatation of the fourth ventricle, hydrocephalus, defects in the skull, as well as ectopia of the bladder and congenital hernias.

The **diagnosis** of spina bifida presents no difficulties when the characteristic picture of the tumor and paralysis are present. In the absence of these concomitant symptoms distinction from lipoma or teratoma may be quite difficult, particularly as these tumors exhibit a preference for the same situation and the association of spina bifida with tumors of this kind is by no means rare. In general it may be said that spina bifida is translucent, only slightly movable, flaccid, covered with a delicate envelope and more or less compressible, the act being sometimes, although not regularly, accompanied by cerebral symptoms. If all these signs, which are not very pronounced, are absent, the presence of spina bifida may be established by means of an exploratory puncture with a delicate acupuncture needle. The X-rays have recently been employed in the examination of a few cases and are useful for the diagnosis of a cleft in the spinal cord, but are of no value for determining the kind of spina bifida that is present.

The **prognosis** in cases of myelocele with pronounced palsies is extremely grave. The ulcerated surface of the sac is prone to become the starting point of a purulent meningitis; if the tumor is very tense it may rupture or at least become permeable for fluid, in that way permitting infection of the interior to take place. Aside from the dangers of intraspinal suppuration, the children may be destroyed by catarrah of the bladder, sepsis following ulcers of the skin, diseases of the intestines, etc. Biedert lost 25 out of 32 cases in the first week. There is no prospect of the palsy disappearing spontaneously; indeed, paralytic symptoms may develop secondarily in cases in which the spinal hernia was at first unaccompanied by symptoms. These palsies are probably due to pressure or to maceration of nerves that pass through a layer of fluid.

Fig. 36.



Spina bifida. Hydrocephalus. Extreme retraction of the head and flexor spasm of the arms due to the hydrocephalus. Marked flexion at the hip-joint (iliopsoas intact). Ulcers on the buttocks caused by the spina bifida.

It is therefore easy to understand that the question of operative removal of spina bifida has for a long time occupied the attention of pediatrists and surgeons. After the first primitive attempts, consisting in clamping or ligation, or compression of the sac, puncture followed by the injection of iodine solutions was resorted to, the injected solution of iodine glycerin being removed soon after its introduction. In this way it was hoped to set up local inflammation and cause adhesions of the inner surfaces of the sac, with complete obliteration. The latest development in this treatment consists in the injection of paraffin saturated with iodine for the double purpose of setting up irritation in the meninges and closing the opening in the spinal marrow with a kind of tampon. At the present time more attention is given to the careful elaboration of operative methods, with the laudable object of avoiding as much as possible any injury to the nerves contained in the spinal sac, although the details of the plastic operations for closing the opening in the spinal marrow are somewhat exaggerated. This is not the place for a discussion of the technic of these operations, for which the reader is referred to text books on Surgery, and to the writings of Bayer, Bockenheimer and others. We shall merely emphasize Bayer's warning against too much operating in cases of this kind. If the patient is completely paralyzed, he is very little benefited by a masterly surgical performance and the substitution of a beautiful scar for a tumor that is constantly exposed to infection; particularly if the paralysis is made worse by the operation, owing to the unintentional removal of large portions of nerves. It also appears that, when there is hydrocephalus, closure of the spinal sac has an unfavorable influence on its subsequent course. Bayer is therefore quite right when he insists on excluding from operation all cases characterized by extreme degrees of paralysis and the presence of hydrocephalus and other pronounced malformations. On the other hand, uncomplicated cases of spina bifida are favorable for operation, and the many statistics which have been collected indicate very good results (Hildebrandt, Nicoll, Sachtleben, Schirmer and others).

A special form of the condition under discussion is called *spina bifida occulta*. In this anomaly, which is usually situated in the lumbar region, there is likewise a cleft in the vertebral column and the spinal marrow is deformed, but there is no accumulation of fluid and therefore no protruding sac. Whether spina bifida occulta is the expression of an intra-uterine process of repair, or, as Hildebrandt no doubt correctly assumes, a special malformation genetically representing the terminal member of the entire series of spinal hernia, is still an open question. The anomaly is characterized by failure of the vertebral canal to close, the occurrence in the spinal canal of fibromuscular tumors due to developmental processes, and by the fact that the spinal marrow is continuous with the external skin (Katzenstein). Externally the malformation manifests itself by a spherical prominence usually situated in the sacral region; sometimes by a depression covered with normal or cicatricial skin and hair such as is not normally present in that situation. Sometimes the cleft in the bone or the above-mentioned muscular tumor can be palpated through the superficial tumor.

The signs of spina bifida occulta are quite remarkable. Congenital palsies are not common and are confined to clubfoot, pes equinus or sensory disturbances. On the other hand, certain more pronounced symptoms, such as incontinence of urine, perforating ulcer, neuralgias and palsies, manifest themselves in the later years of childhood and at the times of puberty. Hence the great majority of reported cases refer to individuals between childhood and adolescence, and the discovery of the condition when the patient is examined usually causes great surprise. I myself know two half-grown boys whose general health is good and who suffer from constantly increasing incontinence of urine, attributable to a spina bifida occulta. Katzenstein has offered a very plausible explanation for the late occurrence of the disturbance; during the normal upward growth of the spinal cord within the vertebral canal, the connecting band between the spinal cord and the skin is subjected to traction, causing distortion of the spinal cord and the gradual development of functional disturbances. Indeed, the removal of such connecting bands has in some cases been followed by disappearance of the disturbances caused by the spina bifida occulta. The operation is therefore distinctly to be recommended as, without such intervention, the symptoms may be expected to increase in severity.

9. *Partial congenital changes of the central nervous system.*—In addition to the congenital disturbances which manifest themselves partly as conspicuous malformations and partly as distinctly recognizable diseases, we find in the nervous system a number of changes affecting a limited area, and which are, according to their seat and extent, of interest to the clinician, or merely of pathologic significance.

(*a*) Brain.—In the brain we have absence of the corpus callosum (Zingerle, Anton, Hochhaus), changes in size of the convolutions (macrogyria and microgyria), and aplasia of the cerebellum. The latter will be referred to again in connection with hereditary ataxia.

(*b*) Of much greater importance are the congenital aplasias in the nuclear region of the medulla oblongata (*infantile nuclear atrophy*). There are certain congenital conditions characterized by complete or partial immobility of the eyes (ophthalmoplegia) absence of facial expression, atrophy of the tongue and interference with its movements, a constant flow of saliva from the mouth, and absence of the lachrymal secretion. These disturbances may be unilateral or bilateral, and the above symptoms may be present only in part. They correspond to paralysis of the ocular muscles, of the facial hypoglossus, etc. Most frequently this congenital motor impairment is limited to the eye and shows itself either in complete immobility of the eye or absence of certain movements, especially in unilateral or bilateral ptosis.

The pathologic foundation in a case of Heubner's was found to be absence of the motor nuclei in the medulla oblongata, namely those of the abducens, facial and hypoglossus. The findings in this case accordingly fully justify the theory of partial lack of development of the central nervous system—infantile nuclear atrophy (Möbius). It seems probable, however, from certain exhaustive investigations by Kunn that the above pathology does not apply to all cases of congenital motor impairment of the eyes and bulbar nerves, and that congenital changes may occur anywhere in the path between the nerve centres controlling the ocular movements and the ocular muscles, and produce motor disturbances. Again, these congenital palsies must be sharply separated from those which develop during childhood. It is thought advisable therefore, to divide all motor defects of the eyes, with or without involvement of the bulbar nerves, into several groups and to distinguish between congenital and acquired changes, between disturbances of nervous, and those of muscular origin. We shall return to the discussion of these conditions after considering muscular palsies.

(*c*) Spinal Cord.—Islands of gray substance are sometimes found within the white medullary tissue, particularly in the posterior columns of the lumbar portion of the cord (also in the medulla oblongata). These anomalies of distribution, however, have no clinical interest and in that respect may be compared to the lateral furrows around the cervical enlargement and around the cord, which are particularly well seen in defects of the pyramidal tract.

There are several varieties of congenital changes of the central canal, which may possibly assume clinical significance. Aside from a moderate local dilatation of the central canal, pear-shaped in cross-section with a long dorsal process ("simple hydromyelia," Zappert), the central canal in newborn infants or somewhat later in life is occasionally greatly dilated, forming a number of diverticula or even completely open at the dorsal wall. This congenital form, in addition to its genetic interest, is significant because it may possibly bear some relation to a subsequent syringomyelia, a theory which finds support in the fact that proliferation of the neurogliar tissue is sometimes observed in the dilated central canal in young infants (and possibly also in the newborn?). The majority of those who have investigated the question of syringomyelia nowadays incline to the view that the condition begins in a congenital defect, which possibly finds expression in congenital hydromyelia and early proliferation of the neurogliar tissue (see Schlesinger's monograph).

These anatomical anomalies occuring during infancy do not give rise to clinical symptoms. If we admit the possibility that many forms of syringomyelia develop from these early changes, it is not surprising that syringomyelia in infancy is practically without clinical symptoms; for it takes some time before the abnormal formations in the spinal marrow attain sufficient extent to produce clinical disturbances. Indefinite prodromal symptoms, not ascertained until later in taking the history of a case of syringomyelia in a youthful subject, may possibly occur during childhood. Except for these, however, syringomyelia is not a children's disease and its discussion in this work seems superfluous.

Occasionally a condition known as *diastematomyelia*, in which the spinal cord is divided into two portions, particularly in the lower dorsal and lumbar regions, is observed at the autopsy table. The development of the two halves of the spinal cord may be approximately equal, or one half may greatly exceed the other in size and development. The rudimentary development of two spinal cords has also been observed. The condition is interesting from a pathological view point only. As a rule it is associated with other malformations of the central nervous system, especially spina bifida.

SECTION II.

ENDOGENOUS DISEASES OF THE CENTRAL NERVOUS SYSTEM

The group of diseases collected under this head contains a number of very different morbid conditions. Their common feature is the occurrence in children, who are healthy at birth, of disturbances in the nervous and muscular systems after the lapse of months, years or decades. The mode of origin of these conditions is generally believed to be that certain portions of the nervous system are endowed with abnormally low resisting power and wear out after a short period of normal functional activity. Gowers describes this condition as *abiotic* atrophy of the central nervous system. The group is also called *endogenous*, *i.e.*, due to internal predisposition, in contradistinction to *exogenous* diseases or such as are due to external causes.

The remote causes of this congenital weakness of certain portions of the nervous and muscular apparatus are not known. Consanguinity of the parents, alcoholism and syphilis or more exactly the so-called parasyphilitic diseases are regarded as exciting causes; for hereditary syphilis which is due to exogenous causes even though operative in utero, does not belong to this group. In many of these endogenous diseases there are very distinct racial factors.

An important feature of the diseases here under discussion is heredity, or the tendency to occur in different members of the same family. All endogenous diseases are encountered either in several generations of the same family or in various members of the same generation (brothers and sisters and cousins). This peculiarity is an extremely valuable symptom, although its absence in an individual case by no means excludes it from the category, since in any hereditary disease some individual must be the first to exhibit the pathological symptoms.

The proof that these conditions deserve to be placed in a special class is furnished by the pathologic findings, which consist in various grades of atrophy degeneration and aplasia; but never, at least in recent cases, acute or chronic inflammations or neoplastic changes.

The number of cases that have been described and classified under this head is practically unlimited. Strictly speaking the group includes not only diseases caused by functional inactivity of the central nervous system, but also diseases in which the muscles, connective tissue, skin, etc., spontaneously undergo morbid changes after a variable period of normal vital activity. According to the special part of the nervous system or muscular apparatus affected and the grouping of the symptoms with relation to one another we have an enormous number of different types, which their observers have attempted to bring into relation with known diseases, as far as possible, under the impression that in finding points of similarity between these apparently inexplicable diseases and tabes or multiple sclerosis an explanation of their pathogenesis would be found at the same time. This attempt to classify the hereditary diseases among exogenous conditions has now been given up. We believe with Jendrassick that the fact that a combination of symptoms cannot be classified according to our conception of the clinical manifestations of cerebrospinal diseases points to the assumption of some endogenous condition. The endeavor to subdivide these diseases into different groups by sharp dividing lines has also been abandoned. It is true that we are sometimes surprised to find that among the endogenous conditions there are certain very characteristic diseases which show a constant and complete similarity to one another; but it is extremely common to find deviations from a type of an otherwise very well known condition, and there are also observed innumerable mixed and transitional forms, which combine the symptoms of one large group of diseases with symptoms of another.

That such a family or hereditary predisposition to some morbid development of a nervous system or parts of a nervous system is conceivable was shown by the investigations of Karpluss on normal brains. He repeatedly observed in various members of the same family peculiar characteristic fissures in the brain, proving that in non-pathologic cases anatomical peculiarities of the nervous system are unquestionably inherited. It is a very obvious conclusion, therefore, that congenitally deformed or morbidly developed portions of the nervous system and consequently also functional disturbances may be inherited in a similar manner.

Why it is that, in spite of the great variety of the endogenous diseases, certain symptoms such as spasticity of the legs, difficult articulation, ocular disturbances, muscular atrophy and the like recur again and again and in similar combinations still remains a mystery. Edinger proposed an ingenious theory of substitution to explain this group of diseases. He assumed that certain portions of the nervous system in these cases lack the power to renew the nerve substances used up in carrying on the functions as is the case in normal conditions.

The following discussion will be limited to the most frequent and well-known combinations of endogenous symptom-complexes, retaining those groups of diseases which formerly were usually described separately. In doing so, however, let it be emphasized once more, that transitional cases between most of these groups also occur.

We offer the following as a provisional classification of endogenous diseases:

A. Spastic Family Affections.
  1. Spinal.
  2. Cerebrospinal symptom-complexes.

B. Hereditary Ataxia (Friedreich's Disease) (Marie's Hérédoataxie cérébelleuse).

C. Muscular Atrophies.
  1. Spinal muscular atrophies.
    (*a*) Cases in adults not occurring in families (including amyotrophic lateral sclerosis).
    (*b*) Infantile spinal muscular atrophy.
  2. Neural form of progressive muscular atrophy.
  3. Dystrophy of the muscles.

D. Bulbar Diseases. Progressive bulbar paralysis.

Appendix.
  (*a*) Myasthenia.
  (*b*) Amaurotic family idiocy.
  (*c*) Thomsen's myotonia.
  (*d*) Periodic paralysis of the extremities

A. SPASTIC FAMILY AFFECTIONS

1. SPINAL SYMPTOM-COMPLEXES

Family spastic paralysis of the spinal type ("hereditary form of spastic spinal paralysis," Erb) is a distinctly hereditary disease which does not always begin in childhood, its first appearance being sometimes delayed until a more mature age. Males seem to be more frequently affected with this disease than females. The symptoms are as follows: after the power of walking is fully established (rarely when the child first attempts to walk) there are noticed a tendency to become easily tired, awkwardness in going up stairs, in getting up from a sitting posture and other similar movements. Gradually the child begins to drag its feet; the feet slip along the floor and the legs are dragged after. The stride is shortened and every step is taken with great caution. Later on, the spasticity becomes more and more distinct, the legs more rigid, tonic tension of the muscles persists, the foot assumes the position of pes equinus, the trunk is bent over forward, and locomotion is possible only with the aid of a cane. Although walking becomes more and more difficult, paresis is not marked in the legs, and the test for gross muscular strength yields a satisfactory result. All the muscles of the legs are in persistent spasm (tension) and do not relax even during rest. The patellar and Achilles tendon reflexes are greatly exaggerated and often of a clonic character; the Babinski reflex is usually positive. Hyperextension of the big toe is often present even without reflex irritation. As a rule the spasticity does not involve the arms. Sensation of every kind is intact; muscle sense and sphincter control are not affected. There is often some diminution of intelligence from the very beginning, and almost regularly later in the course of the patient's life.

The course of hereditary spastic spinal paralysis is extremely tedious. The patients do not become bedridden, or only very late, and the disease does not shorten the duration of life.

The few cases that have come to autopsy show the pathological picture of a simple system-disease, with sclerosis of the pyramidal tracts. The lateral cerebellar tract and the columns of Goll showed slight changes. The brain is intact.

A form, which, although somewhat different, nevertheless belongs to this group, was described by Jendrassick, who was able to demonstrate as the cause of the spastic phenomena in the legs a shortening of the muscles and tendons.

2. CEREBROSPINAL SYMPTOM-COMPLEXES

The characteristic signs of these forms of endogenous diseases are nystagmus, strabismus, disturbances of speech (bradylalie) deficiency of the intellect, spasm (spasticity), and palsies of the legs and arms. Other symptoms that occur with varying regularity are: atrophy of the optic nerve, ocular palsies, tremors, wobbling of the head, athetosis, disturbances of deglutition and other bulbar symptoms, isolated palsies, weakness of the sphincters and finally—indicating a transition to the myopathies,—muscular atrophy and pseudohypertrophy. The grouping of these multifarious morbid symptoms in an individual case or individual families is subject to wide variations; hence the publication of numerous interesting cases belonging to this category and the erection of a series of different types of familial palsies, which, as has been shown above, cannot be regarded as nosologic entities.

The onset of the disease, or, in other words, the manifestation of the first symptoms may occur at any age; most commonly children beyond the tenth year of life are attacked. There are also cases, however, in which the first symptoms make their appearance very early, so that the dividing line between congenital and endogenous conditions cannot be sharply drawn. Boys as well as girls are attacked, sometimes in the same family, and the disease may be transmitted through the mother as well as through the father. Consanguinity appears to play an important rôle. Sporadic cases without any demonstrable hereditary or family elements are rare in this form of endogenous disease, or else they are not recognized as cases of cerebrospinal spastic paralysis.

The initial **symptoms** are extremely variable. Among the earliest are spasms in the legs, which interfere with walking and lead to contractures. At the same time, or a little later, nystagmus and slow speech make their appearance. Still later, awkwardness of the hands, tremors, choreic and ataxic movements, and often wobbling of the entire body are added to the picture. The intelligence suffers later in the course of the disease. Mingled with these cardinal symptoms of family spastic paralysis, the manifestations already mentioned, particularly ocular palsies and bulbar symptoms (disturbances of deglutition, dribbling of saliva, forced laughter, etc.) are frequently encountered. Trophic changes in the skin and in the osseous system have been very rarely observed.

Special attention should be called to certain symptoms of the cerebrospinal family diseases because they suggest other forms of endogenous diseases, or represent intermediate forms between the two. We refer to cases with disturbances of coördination, as in hereditary ataxia, with muscular atrophies of the spinal type and, finally, pseudohypertrophy of individual muscles. Such cases show very clearly the impossibility of delineating definite clinical pictures in the familial spastic palsies.

Among the numerous cases belonging to this category those reported by Pelizæus, Freud, Krafft-Ebing, Wagner, Luce, Bruns, Gee, Pribram, Dreschfeld, Haushalter, Bouchard, Homen, Ganghofner, Jendrassik, Spiller, Rolly, and Oppenheim are deserving of special mention.

The **clinical course** in diseases belonging to this group is slowly progressive, and the disease does not materially interfere with the patient's comfort. When, however, he is confined to bed on account of the increasing motor disturbances, when dysphagia and loss of bladder control become pronounced, a fatal termination may at any time be brought about by direct sequelæ of the disease such as bedsores, pneumonia or cystitis. This slowly progressive character of the disease, interrupted by periods of temporary arrest of the symptoms, is characteristic of this group of nervous conditions and, when a case is under observation for some time, assumes a diagnostic importance. Next to the progressiveness, an hereditary or family tendency is significant in a diagnostic sense. These two are the only reliable diagnostic aids, and in the absence of both, or if they cannot be elicited with certainty, the differential diagnosis from cerebral infantile palsy, brain syphilis or neoplasm is difficult.

Post-mortem examination of these cases reveals some remarkable findings. Degeneration of extensive segments of the spinal tract, particularly the pyramidal tracts, the lateral cerebellar tracts and the posterior columns is found. These pathologic findings are described as system-diseases, and we speak of a combined system-disease when, as in this instance, several systems of nerve tracts in the spinal cord are affected (Kahler-Pick, Westphal, Strumpell). Compared with these changes due to a congenital defect others, such as local inflammation of the meninges, cellular degeneration, and the like, are of minor importance. The question whether the degenerations are primary, or secondary to other endogenous diseases, is still under discussion, and its decision is rendered more difficult by the fact that most of the cases that come to autopsy are of long standing—years or decades—so that abundant opportunities must have been present for the development of secondary changes in the central nervous system. In the case of the combined system-diseases which occur in this group of family affections—for some occur after intoxication and other diseases—the degeneration is probably regarded by most neurologists as primary in character.

B. HEREDITARY ATAXIA

In the year 1861 the Heidelberg clinician, Friedreich, described a symptom-complex which occurs in members of the same family, and is characterized by onset during childhood, ataxia, absence of the patellar reflex, nystagmus, pes equinus, and a slowly progressive course. Schultze later had an opportunity of examining a case post mortem and found changes in the posterior columns of the spinal cord which he regarded as the pathologic foundation of the disease. The existence of this clinical picture has since been confirmed by an extraordinary number of publications; a Frenchman (Brousse) gave it the name of Friedreich's disease. Gradually, however, it was found that the disease was less sharply defined than it had at first been supposed. It was observed in older individuals; some of the characteristic symptoms were found to be absent; and finally, post-mortem examination of a few cases revealed other disturbances of the central nervous system such as degeneration of the pyramidal tracts and changes in the cerebellum. After Senator had been led by the pathologic findings to question our original conception of the disease, Marie came forward with the description of a morbid condition which, along with many points of resemblance to Friedreich's disease, exhibited so many peculiar features that he deemed it necessary to regard it as an independent nosologic entity. Clinically this condition differed from Friedreich's disease by its onset at a later period of life (after the age of 20), greater uncertainty in the gait (cerebellar gait), the fact that tendon reflexes were present or exaggerated, ocular palsies and, occasionally, atrophy of the optic nerve. Post mortem, hypoplasia of the cerebellum with secondary degenerations were found. Marie gave it the name of hérédoataxie cérébelleuse. The existence of this disease was also soon confirmed by numerous cases reported in the literature (Londe's collection). It seemed as if a distinct advance had been made in the differentiation of the individual forms of hereditary ataxia, but this distinction also was found to be untenable. Cases were soon discovered which combined symptoms of both diseases. Then came the reports of autopsies with simultaneous changes in the cerebellum and in the spinal cord, and others with atrophy of the cerebellum but without cerebellar symptoms. In view of these facts most authors (first of all Londe, then Bäumlin, Seiffer, Veraguth, Oppenheim, Raymond and quite recently Nonne and others) have expressed the opinion that Friedreich's tabes and hérédoataxie cérébelleuse can no longer be distinguished either clinically or pathologically and that it is more correct to include all these conditions under the head of hereditary ataxia, with preponderance of the spinal symptoms in some cases and of the cerebellar symptoms in others. Between ataxic diseases and other forms of endogenous diseases even the dividing line has been shifted since Bäumlin and others described cases of Friedreich's disease associated with muscular changes.

Hereditary ataxia is a distinctly hereditary disease and has been observed in two or three successive generations. As a rule either the male members or the female members of a family are exclusively attacked, but there appears to be no difference between the sexes as regards frequency. Isolated cases of the disease are not rare and are usually of the spinal form; the fact that single cases are frequently observed in France has been explained with some apparent reason by Socas as due to the small number of children in that country. Whether in this form of hereditary disease consanguinity and alcoholism in the parents, infectious diseases and traumatism play an important part is doubtful and indeed very unlikely as regards the last two factors. Edinger's theory, so far as it applies in Friedreich's tabes, has been exhaustively discussed by Bing.

In the spinal form the onset of an hereditary ataxia occurs in childhood usually between the ages of four and seven (Rütimeyer's case in a child two years old, Seiffer). Vizioli's analysis gives the onset during the first decade of life in 39 out of 60 cases, but exceptions also occur in which the disease begins at a later period. In cerebellar ataxia the first symptoms, according to Marie and others, appear during the twenties and according to other authors at a still later age; but there are also cases of Friedreich's ataxia with very early onset (Fraser, Bäumlin and my case in a child three and a half years old). It appears therefore that there is no constant difference between spinal and cerebellar forms of ataxia as regards the age when the disease first makes its appearance.

The first and characteristic **symptom** of ataxia is uncertainty of the gait. The child walks with legs wide apart and the body sways from side to side. In severe cases, particularly when the cerebellar symptoms predominate, the gait becomes reeling like that of a drunkard (cerebellar gait). The disturbance of coördination affects not only the walking (locomotor ataxia) but the standing and sitting postures as well (static ataxia), manifesting itself in a constant search after fresh points of support and inability to keep quiet. Romberg's phenomenon (swaying when the eyes are closed) is less marked in the spinal than in the cerebellar form. The motor unrest is not confined to the legs and early involves other portions of the body, producing wobbling of the head, tremor of the hands, resembling intention tremor, and of the entire trunk, at times exhibiting the character of a simple tremor, at others that of choreic or athetoid movements. Pronounced partial or complete paralysis of the legs is rare and, with few exceptions, constitutes a late symptom of hereditary ataxia. On the other hand, peculiar malformations of the feet are quite characteristic of the spinal form of ataxia. The malformations are caused by overextension of the large toe, which may be an early symptom. Rütimeyer reports that in a certain family the anxious parents recognized by this symptom alone that another of their children had been attacked by the baneful disease. Sometimes the deformity does not proceed beyond this hyperextension of the great toe, with pes equinus. As a rule, however, it is followed by a further malformation of the foot and clawlike position of the toes. Whether these changes in the foot are due to atrophy of the tarsal bones of the foot (Duchenne and others) or to disturbances of equilibrium (Besold) is still undecided. The Röntgen rays do not show any anomalies in the skeleton. This deformity of the foot, which was formerly ascribed exclusively to Friedreich's disease, has also been observed in cases which otherwise correspond to the cerebellar type.

In regard to the behavior of the patellar reflex the difference at first observed between the two forms of ataxia has also been found to be inconstant. It is quite true, however, that the reflexes usually disappear early in Friedreich's tabes and are exaggerated in hereditary ataxia; but there are spinal forms with exaggerated, and cerebellar forms with diminished reflexes, and the reflexes may differ in the two legs or in members of the same family who otherwise present the same disturbances. A frequent concomitant of spinal ataxia, when it has existed for some time, is scoliosis or kyphoscoliosis, which must be attributed to weakness of the spinal muscles. Marked muscular atrophy occurs both as the result of disuse, particularly in the legs, and in the form of isolated muscular atrophies, particularly of the muscles of the hand. Combinations of hereditary ataxia with atrophy of the peroneal type or pseudohypertrophy form the transition to the myopathies, which will be described later.

Sensory disturbances do not belong to the regular picture of hereditary ataxia, although they have been observed, especially in the cerebellar form. The muscle sense is always intact. Spontaneous pain, particularly pain of lancinating character, is extremely rare. The cutaneous reflexes are usually preserved but sometimes diminish in strength in the course of the disease. Paralysis of the bladder and rectum develops only in very advanced cases.

Hereditary ataxia is accompanied by a series of very important cerebral symptoms. Nystagmus is extremely common in the spinal form and usually absent in the cerebellar type. Disturbances of the ocular muscles, such as strabismus, ptosis, and diplopia, have frequently been observed, especially in Marie's form. Geniune or neuritic atrophy of the optic nerve is quite common, particularly in hérédoataxie. The pupils are usually normal. Disturbances of speech are regularly present in hereditary ataxia. Speech is slow, awkward and sometimes scanning. Marie compares it to the cerebellar gait. Disturbances of speech are particularly associated with Friedrich's form of ataxia and increase in severity as the disease progresses. The intellect is also impaired particularly in the spinal form, and there may be actual dementia. Among the unimportant and infrequent symptoms are vertigo, disturbances of the respiration, profuse salivation and forced laughter.

The **course** of hereditary ataxia is fortunately extremely slow. The malady progresses very gradually and it is usually years before the patient is confined to his bed. Even after that, a period of from five to ten years may elapse before life is terminated by some intercurrent disease, usually some affection of the respiratory organs.

The **pathologic findings** in hereditary ataxia, based on a large number of autopsies, also represent numerous transitions between what is found in typical cases of Friedreich's tabes and the pathology of hérédoataxie cérébelleuse. The characteristic changes in Friedreich's ataxia are diminution of the spinal cord as a whole, sclerosis of the posterior columns, that is, the columns of Goll in their entire extent and the columns of Burdach, in varying degrees of intensity and extent—in short, disease of the posterior roots. In addition, other portions of the spinal cord, such as the lateral cerebellar tract, the columns of Clark and Gowers' bundles are usually affected. Friedreich's disease is therefore an example of a system-disease—simple when the posterior columns only are diseased and combined when the pyramidal tracts also are affected. In typical cases of hérédoataxie cérébelleuse marked atrophy of the cerebellum as a whole and of its cellular and nervous elements, and degeneration of the nerve tracts derived from the cerebellum are found. This condition, which used to be regarded as characteristic of the cerebellar form, is not confined to this type, since it has been found in cases of Friedreich's simple tabes; while conversely, these cerebellar symptoms may be absent in apparently typical cases of Marie's disease. If we also include under the head of Marie's disease cases with simultaneous lesions in the cerebellum and in the posterior columns, we see that the uncertainty of the clinical distinction between the two forms finds its counterpart in the pathologic picture. We may therefore regard hereditary ataxia as an endogenous disease of the static system, in which we must include both the organ of equilibrium (cerebellum) and the systems of the posterior columns.

The **differential diagnosis** from tabes dorsalis and other syphilitic affections, brain tumor, and the spastic diseases may present some difficulties. But ataxia as a rule is not an early symptom in infantile tabes, while, on the other hand, rigidity of the pupils, bladder disturbances and lancinating pain are frequent. A brain tumor, particularly when situated in the cerebellum, often produces symptoms simulating those of hereditary ataxia; but the signs of general cerebral pressure are more pronounced and the course is more rapid. A sharp distinction from the other forms of hereditary disease is not of very great importance since the occurrence of transitional forms is quite characteristic of these hereditary conditions. Hysteria, neurasthenia and familial tremors rarely need to be considered in the differential diagnosis.

C. MUSCULAR ATROPHIES

It has been known since about the third decade of the last century that there occur in adults and children diseases consisting in atrophy or increase in volume of the muscles, with paralysis of a progressive character. Gradually, through the investigations of Aran, Duchenne, Erb, Schultze, Leyden, Moebius, Charcot, J. Hoffman and many others, a number of different forms of these muscular atrophies were distinguished, and this led to the erection of different types and incidentally to the subdivision of all the progressive muscular atrophies into a number of subgroups. The basis for this detailed classification was found in the clinical course, the age when the disease makes its appearance, the tendency to occur in members of the same family and, especially, in the pathology. On pathologic grounds the division was made into spinal and myogenous, according as the motor cells of the spinal cord or the peripheral muscular system were found to exhibit pathologic changes. It was also found that the spinal form as a rule attacks adults and does not tend to occur in members of the same family, whereas the myogenous variety preferably occurs in children and exhibits a family tendency. Thus, the different kinds of muscular atrophy appeared to be sharply differentiated, but before long this sharp division had to be abandoned. On the one hand, Werdnig and Hoffmann described a pronounced spinal and familial disease of the muscles in earliest childhood; on the other hand, isolated cases were observed presenting signs of both forms of the disease, and occasionally pathologic changes in the spinal cord were found in apparently myogenous forms of muscular atrophy. Thus the sharp line of distinction between spinal and myogenous atrophy became somewhat blurred, and with regard to the individual varities of myogenous muscular atrophy the finding of certain constantly recurring complexes had led to the classification of certain definite and distinctive types. But, as in the case of the forms of diseases occurring in families which we have described above, more and more "impure cases" and "transitional forms" were observed, making it impossible to retain the classification into the usual types. Erb deserves the credit of having spoken the decisive word on this question by grouping all myogenous diseases under the general head of muscular dystrophy, and at present most authorities have accepted this standpoint. Hoffmann's neural (neurotic) atrophy represents an apparent transitional form between spinal and muscular paralysis of muscles. With this reservation we shall in the following discussion retain the customary classification of muscular atrophies, as it will best enable us to sift the large amount of available material.

I. SPINAL MUSCULAR ATROPHY

(a) *Forms in Adults not Occurring in Families*

The fact that this disease only exceptionally attacks (larger) children seems to make a detailed description of it superfluous in this work. It has never been definitely proved that these muscular atrophies are endogenous in character. The symptoms of the disease are quite typical: First, atrophy of the muscles of the hand (thenar and antithenar [hypothenar]). Extension of the atrophy to the muscles of the shoulder, neck and throat and, finally, atrophy of a lesser degree in the legs. All the affected muscles are flaccid and atonic, and function is soon completely abolished. The tendon reflexes are diminished or absent; electrical irritability greatly impaired. The paralysis is often preceded by fibrillar twitching. Sometimes the muscles of the shoulder-girdle are primarily attacked. The course is slow and may extend over years. Death results, if not through some intercurrent disease, from involvment of the bulbar nerve centres and paralysis of the diaphragm. Postmortem a pronounced atrophy and degeneration of the motor cells of the spinal cord, the root fibres and peripheral nerves, and severe atrophy and degeneration of the muscle substance are found. So called amyotrophic lateral sclerosis represents a special form of spinal muscular atrophy. It is only exceptionally a disease of childhood, although there is some reason to assume a congenital predisposition. In the distribution of the palsies, their extent, and in the presence of fibrillary twitching it presents much similarity to spinal muscular atrophy; but, while there is atrophy of the arm muscles, there is weakness of the legs accompanied by spasticity and great exaggeration of the reflexes, which are also preserved for some time in the arms. The course is more rapid than that of the simple spinal form and the occurrence of terminal bulbar phenomena quite frequent. Pathologically the two forms are distinguished by the fact that the pyramidal tracts are also degenerated in amyotrophic lateral sclerosis, which is therefore a disease of the entire cortico-muscular nervous apparatus.

(*b*) *Early Infantile Spinal Progressive Muscular Atrophy*

This was first described by Werdnig, and later a well-defined clinical picture was outlined by Hoffmann in a series of monographs. The symptomatology of this disease is a valuable addition to our knowledge. It is a disease of the first years of life and exhibits a marked tendency to occur in members of the same family. Almost all the cases that have been described, about 30 in number, occurred in several children of the same family. The disease could be traced through two or three generations. The *symptomatology* is as follows: A previously healthy child in the second half of its first year develops weakness of the legs, the back, then of the muscles of the neck and shoulder and finally of the hands and toes, so that in a short time it becomes completely paralyzed. The affected muscles exhibit marked atrophy, rarely a pseudohypertrophy. Fibrillary twitching is not constant; the reflexes soon disappear and there is marked disturbance of the electric irritability of both nerves and muscles. The cranial nerves and the sphincters escape; speech and the sensorium are not affected. There is no pain. Death ensues in from one to four years from involvement of the muscles of respiration, pneumonia, or intestinal catarrh. Atypical cases, both as regards the period of onset and the duration, have been described (Beevor, Bruns and Torild). The pathologic basis of the disease is a uniform atrophy of most of the cells of the anterior horns in the entire spinal cord, degeneration of the root-fibres and of the motor nerves, and pronounced muscular atrophy. Prognosis and treatment are hopeless.

2. NEURAL FORM OF PROGRESSIVE MUSCULAR ATROPHY (PERONEAL TYPE)

This condition, which is peculiar to childhood, has been known for a long time (Eulenberg, 1856), but its accurate definition we owe to Charcot, Marie, Tooth and, particularly, Hoffmann and Bernhardt. Hoffmann originated the term progressive neurotic (or neural) muscular atrophy, while Bernhardt called the disease spinal neuritic atrophy. It is distinctly hereditary or rather familial, and occurs chiefly in later childhood, somewhat more frequent in boys than in girls. The atrophy appears first in the peroneal muscles or sometimes in the other muscle groups of the legs. There results a peculiar disturbance of the gait (stepping gait). The foot is raised very high, the toes are extended and point downward, and are the first to come in contact with the ground as the foot is put down. Sometimes a similar disease appears at the same time in the arms, particularly in the muscles of the hands and in the extensors, producing the deformity known as claw-hand. The paralysis sometimes extends to other muscles of the extremities, but the rest of the muscular system usually escapes. The patellar and Achilles tendon reflexes diminish as the disease goes on and usually disappear altogether. Fibrillary twitching in the diseased muscles or those about to be attacked is quite frequent. The electric irritability of the atrophic muscles is variable. As a rule the reactions of degeneration are present, or there is complete absence of reaction to either the galvanic or faradic current. Sometimes muscles that are still perfectly capable of functionating, and are not atrophic, exhibit marked changes in their electric behavior. Sensory disturbances are not rare in neural muscular atrophy and manifest themselves in attacks of pain, hyperæsthesia, or diminution of the tactile and pain sense. Vasomotor phenomena have also been observed. The disease is extremely slow and often does not progress for years beyond paralysis of the legs. The progress of the disease may be interrupted at any time by remissions and exacerbations (according to Goldenberg always in the spring).

The **pathology** of the progressive muscular atrophy has never been fully cleared up. According to Hoffmann's view the peripheral nerves appear to take an active part in the disease process, for they are found to be distinctly degenerated. But the spinal cord also shows changes which can be attributed only in part to the primary nervous disease. Hence, it is at least a question whether both spinal or neural changes may not take part simultaneously in the pathology of this disease. For this reason Bernhard designated the disease "spinal neuritic atrophy." If we include the cases in which a primary muscular change must be assumed (myositic form of progressive muscular atrophy), we have sufficient proof that the original expectation of finding in this form of muscular atrophy a disease which should form a neurogenic connecting link between the myelogenic and the myogenic forms has not been realized.

A deviation from the normal type of this disease is found in a condition first described by Déjérine and Sottas and carefully studied by Brasch. In the main this condition resembles the above-described form of the disease, with the addition of pupillary rigidity, miosis, Romberg's sign, ataxia and other symptoms. The two French authors demonstrated both clinically and pathologically a thickening of the nerve trunks in this disease, which they accordingly named interstitial hypertrophic progressive neuritis. But the pathology of these cases is still in doubt, and most authors include them under the head of neural muscular artophy, the association of symptoms belonging to hereditary ataxia being a feature of great interest.

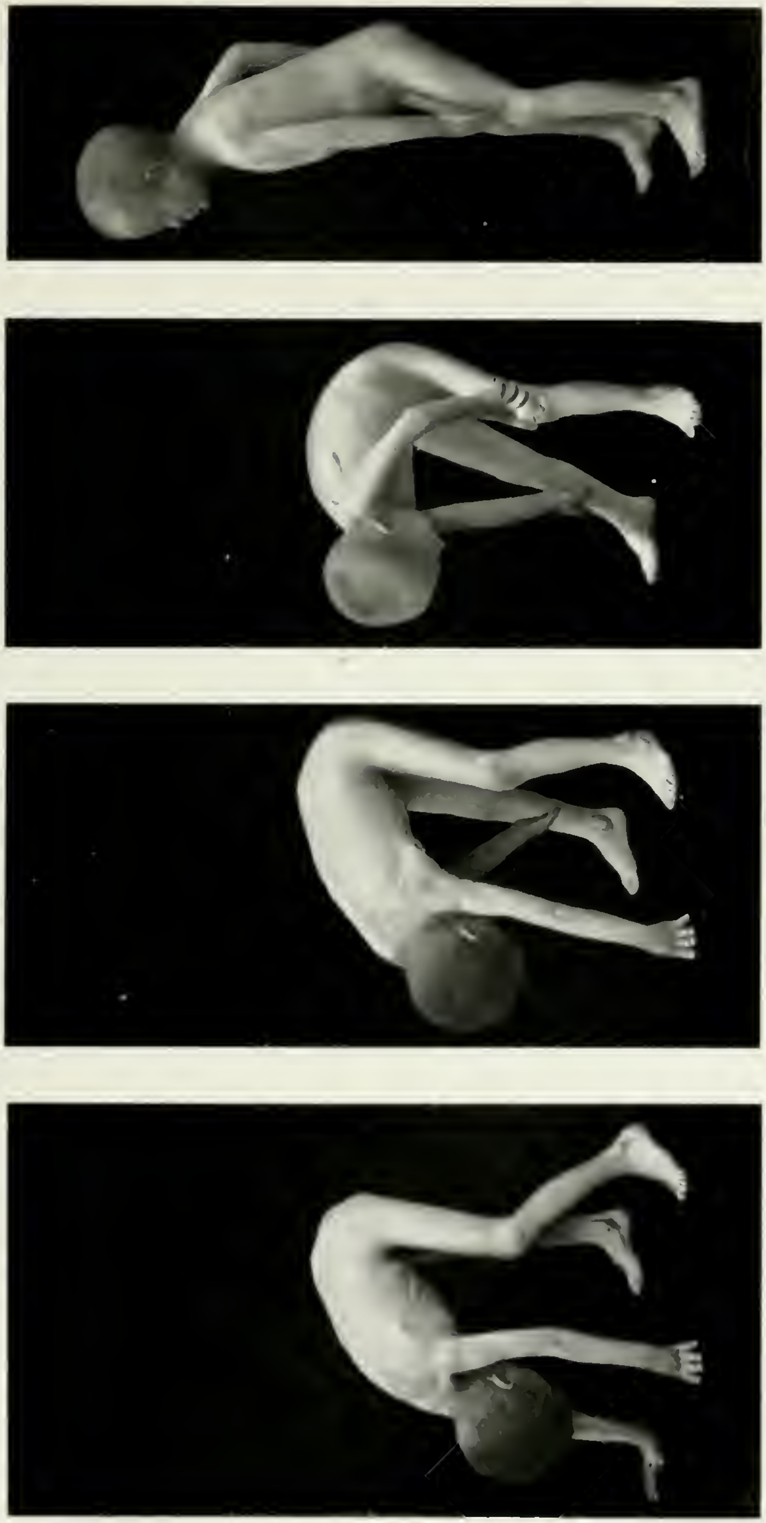
3. MUSCULAR DYSTROPHY

In this group we include, after Erb, Raymond and most neurologists, a number of muscular atrophies which were described by their first observers as special types of myopathies and accordingly regarded as so many different diseases of the muscular system. Depending on the time of appearance, the localization of the first symptoms, the character of the course, and the predominance of atrophic or pseudohypertrophic changes in the muscles, a number of different forms were distinguished, which are described in the literature as the *juvenile form of Erb;* the Landouzy-Déjérine *infantile type* (*facio-scapulo-humeral*); the *hereditary* form of Leyden and Möbius, and *pseudohypertrophy.* The characteristic feature of muscular dystrophy is that the spinal cord escapes; although transitional forms between simple muscular dystrophy and spinal muscular atrophy have also been described.

The disease is usually transmitted by the mother. Boys are affected somewhat more frequently than girls. Members of the same family usually present the same type of dystrophy. With the exception of family predisposition, all other etiologic factors that have been advanced, such as exposure to cold, traumatism and infectious diseases have not even the dignity of an occasional cause. Babinski and Onanoff made the interesting observation that the disease simultaneously attacks those muscle groups which develop at the same time in the embryonal structure.

The *onset* of muscular dystrophy usually occurs in childhood, exceptionally during the adolescent period. Occasionally it is learned that the children have always been awkward, and have learned to walk with difficulty. The progress of all the different forms of dystrophy is exceedingly slow, slower than that of the spinal muscular diseases. The patients may live for years and even decades, but they spend

PLATE 60.



MUSCULAR DYSTROPHY.
Paresis of muscles of back. "Climbing up one's self" in four phases.

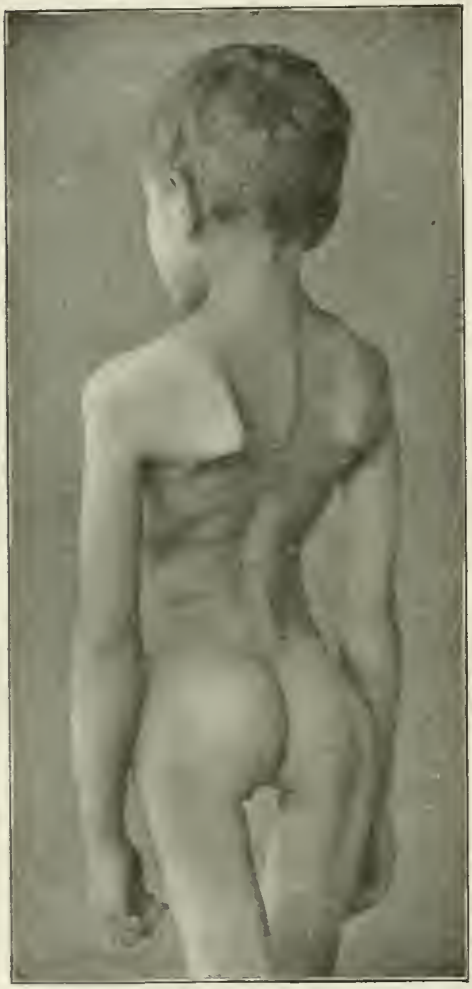
most of their dreary existence in bed. Death ensues from intercurrent diseases, particularly tuberculosis.

The muscular affection in dystrophy presents the following peculiarities: (1) The disease manifests itself in atrophy, temporary hypertrophy and pseudohypertrophy (increase in the volume of the muscle from proliferation of the connective tissue attended by atrophy of the muscular elements). (2) The individual muscles are not affected uniformly; the morbid process presents a disseminated character and not until later extends to the entire muscle. (3) Fibrillary muscular tremors are usually wanting. (4) While the electric irritability of the muscles is often diminished so that they sometimes respond sluggishly, the usual law of reaction is not reversed nor is electric irritability abolished except during the terminal conditions. (5) The paralysis is usually flaccid; but, in contradistinction to a spinal palsy, the muscular tone is often present. Contractures sometimes develop, especially in the foot. (6) The tendon reflexes are preserved for a long time and are diminished only in palsies of long standing. (7) The muscle fibres exhibit atrophy and hypertrophy, as well as increase of the muscle nuclei and fatty degeneration of the individual muscle fibres. Macroscopically the muscles present a peculiar pale red or yellowish color.

Degeneration of the cells of the spinal cord and of the anterior roots has been observed particularly in cases of long standing, but it is not improbable that these changes may be secondary. Nevertheless, this degeneration, and the fact that cases of spinal atrophy combined with pseudohypertrophy of the muscles occur, indicate that the distinction between the myelopathic and myopathic muscular diseases cannot be sharply drawn.

The individual types of dystrophy present the following features: (1) In the *juvenile form of Erb*, which preferably occurs during late childhood and the years of puberty, the atrophy begins in the shoulder girdle; the arm and the hands escape. Later in the course of the disease the muscles of the pelvic girdle, of the back and of the thigh, rarely the abdominal muscles are affected. True and false hypertrophy is observed both at the beginning, and during the later course of the disease. (2) The *infantile* or *facio-scapulo-humeral form* (Landouzy-Déjérine) manifests itself first in the face, in the sphincter muscles of the eye and mouth, and in the masseter. The face assumes a mask-like appearance, the play of features is lost, and there is inability to close the eyes and round the lips. Pseudohypertrophy sometimes develops in the muscles of the face, affecting particularly the lips, which become thickened and unsightly (tapir lips). Later the muscles of the shoulder and arms are attacked. (3) The so-called *hereditary form* first shows itself in weakness of the muscles of the back, lordosis, and difficulty in walking, without pseudohypertrophy. This form of dystrophy preferably attacks children from eight to ten years of age. (4) *Pseudohypertrophy*, the most frequent and best known form of dystrophy, first attacks the long muscles of the back, the calf muscles and the extensor muscles of the thighs. Pseudohypertrophy develops early in the calf muscles and soon afterwards in the diseased muscles of the thigh, causing a very striking and characteristic appearance which cannot well be mistaken. Later in the course of the disease the muscles of the shoulder, arm and lumbar region are affected, the deltoid and supraspinatus and infraspinatus muscles showing a tendency to pseudohypertrophy. The forearm and hand escape. Pes equinus and contractures in the calf muscles are not rare. These palsies first produce a peculiar waddling or rocking gait and there is marked lordosis of the sacral region. The children are unable to raise the trunk quickly from the stooping or recumbent position and help themselves by bracing their hands against their own legs "they climb up on themselves" (see Plate 60). In the beginning of the disease they are sometimes able to raise themselves by a sudden jerk. The difficulty in walking shows itself particularly in going upstairs, in greatly diminished resistance to fatigue, and in a tendency to fall over. The weakness of the shoulder muscles is recognized by defective fixation of the shoulders when one tries to raise the child with its arms pressed against its sides. When the child tries to raise the arm, an effort to find support along the back of the neck and head is also noted. (The hands climb up along the head.) Partial paralysis of the abdominal muscles causes arching of the abdomen and interferes with abdominal pressure. The abdominal reflex is absent in these cases. If the legs, the shoulder girdle, the muscles of the back and the pelvic girdle are paralyzed, the patients become absolutely helpless,

Fig. 37.

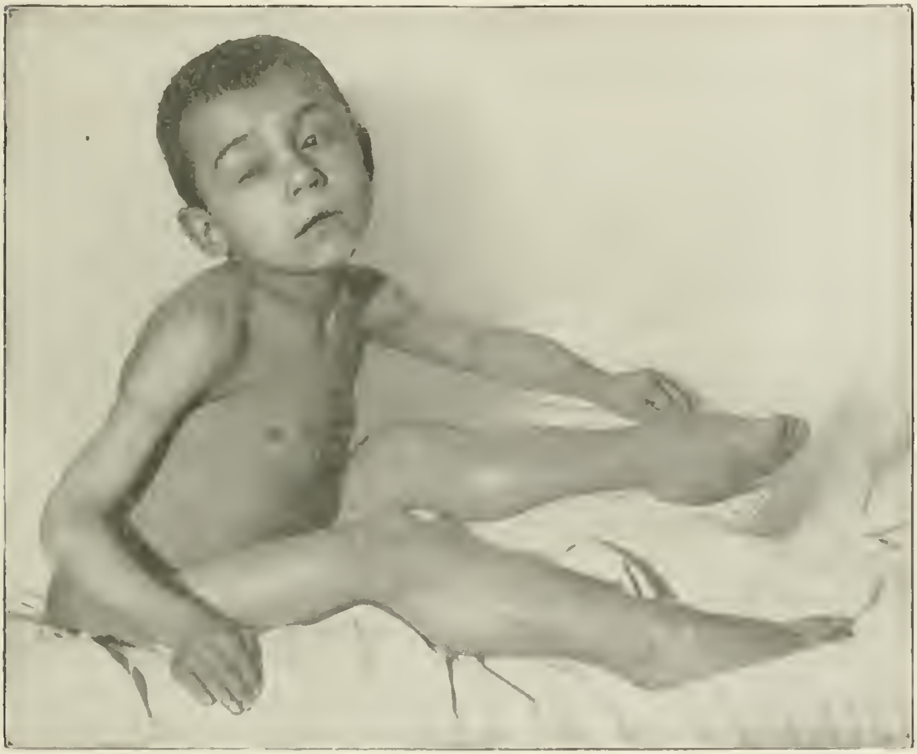


Muscular dystrophy ("hereditary form"). Atrophy of the muscles of the back, lordosis, atrophy of the thigh muscles.

and their condition offers a strange contrast to the athletic development of the body which may be simulated by the pseudohypertrophy. The muscles of the face almost always escape in pseudohypertrophy. Involvement of the eye movements is very rare. The sphincters in these, as in all other forms of dystrophy, remain intact. In advanced cases the intelligence sometimes suffers and speech is disturbed.

Differential diagnosis.—Muscular dystrophy must be differentiated from the spinal form of muscular atrophy and from the neurotic form. Characteristic features of muscular dystrophy are the failure to involve the hands and forearm, the absence of fibrillary twitching, the preservation of the patellar reflexes which are usually normal, and the presence of pseudohypertrophy. From syringomyelia, a disease which rarely needs to be considered in children, muscular dystrophy is distinguished by the fact that the hands escape and by the absence of severe sensory disturbances. Rachitic disturbances of the gait, congenital dislocation of the hip, beginning spondylitis, and spinal meningitis may be confounded with dystrophy. But the simple precaution of bearing these things in mind will enable one to arrive at the correct diagnosis after some observation of the case.

FIG. 38.



Muscular dystrophy (pseudohypertrophic form). Boy nine and a half years old. Marked thickening of the calf muscles. Pes equinus; paresis of the legs.

D. BULBAR DISEASES

In the discussion of congenital diseases we referred to conditions characterized by the presence of ocular palsies and, more rarely, of other cranial nerves, and in accordance with the autopsy findings of Heubner we ascribed these conditions to a congenital aplasia of the nuclear region. Similar palsies of cranial nerves may also develop in earliest childhood, due most probably to hereditary or familial causes. Palsies of this kind, when affecting the ocular muscles, were described by Möbius among the congenital conditions and his description of the clinical picture of infantile nuclear atrophy was made to include these cases also. This classification, however, was given up by later authors (Kunn and especially Peritz). According to these authors the nuclei of the cranial nerves are subject to the same abiotic processes as the motor cells of the spinal cord, and in that case certain forms of ophthalmoplegia and bulbar symptoms entirely analagous to spinal muscular atrophy may develop, which either become arrested at a certain stage or have a tendency to progress indefinitely. We have autopsy reports (Heubner and Naef) which confirm the existence of this medullary form of endogenous cranial nerve palsies and which definitely prove the occurrence of atrophy of the corresponding nuclei. Clinically also we observe intermediate forms between spinal and bulbar diseases; thus, spinal palsies, on the one hand, may lead to bulbar diseases and on the other hand, cases beginning with bulbar symptoms may later develop paralysis of the skeletal muscles.

1. *Infantile progressive bulbar paralysis* is often observed in brothers and sisters. The first symptoms usually appear late (from the 6th to the 10th year of life) and consist in disturbances of speech and deglutition, sluggishness of the muscles of expression, dribbling of saliva and failure of the lachrymal secretion. Later on atrophy and tremor of the tongue, a mask-like expression of the face, paralysis of the soft palate, hoarseness, weakness of the muscles of mastication, irregularity of the respiration and of the pulse are superadded, and the child usually dies. Occasionally the disease remains stationary for some time.

2. *Progressive ophthalmoplegia* affects either all the ocular muscles or only individual ones, especially the levator palpebræ superioris and one abducens. The internal muscles of the eye always escape. Combinations with palsies of other cranial nerves (facial) are not rare. The ultimate effect of these disturbances of the ocular muscles is permanent interference with the movements of the eye, but life is not endangered.

Although these disturbances, which are due to a nuclear lesion, are analogous to the spinal diseases, it may possibly be justifiable with Kunn to regard disturbances of the ocular muscles occurring early and associated with changes in the peripheral eye muscles as dystrophic processes. The subject has been very little studied and further investigations would be acceptable.

APPENDIX

(a) *Pseudoparalytic Myasthenia*

The investigations of a number of authors in recent years have led to the recognition of a clinical picture, the pathology of which is still unknown, but which is probably to be included among the endogenous diseases (possibly a primary muscular affection).

The characteristic feature of the disease is a weakness of the bulbar muscles, including the ocular muscles, manifesting itself in gradual loss of function and particularly in greatly diminished resistance to fatigue. Thus, the individual may be able to close the eyes, to speak, or, in severe cases, to swallow once or twice; but he is unable to repeat the acts and several minutes must elapse before he is again able to do so. The muscles of the trunk, extremity and back of the neck exhibit a similar behavior. The muscles are not atrophic and there are no reactions of degeneration; but irritation with the faradic current in a very short time fails to produce a contraction, and a short period of rest is required before the contraction again takes place (myasthenic reaction). So far the disease has been observed chiefly in young individuals, but not in children. Pathological examination of the nervous system yields absolutely negative results, but quite recently certain inconstant changes in the muscles (cell accumulations) have been found which are difficult to interpret. Curiously, patients with this disease are sometimes the subjects of other grave affections, particularly neoplasms in the mediastinum (with persistent thymus), suggesting the possibility that the injury to the muscles may be due to the action of some morbid metabolic products. But as, on the other hand, congenital disturbances are sometimes associated with myasthenia, opinions incline to the theory of a congenital defect capable, under certain conditions, of causing the disease. The prognosis is very grave although the disease may become arrested and show no tendency to progress for some time and apparent improvement may take place, most of the cases terminate fatally from dyspnœa, the entrance of food into the windpipe, asphyxia or inanition.

(b) *Amaurotic Family Idiocy*

Amaurotic family idiocy occupies a peculiar position among the familial diseases. The hereditary character of the disease, age of the patient, and the symptomatology are so characteristic as to distinguish it from most other diseases occurring in families and make it in reality a sharply defined nosologic entity. It is quite true that our knowledge of the disease is still comparatively recent, and for that reason it is possible that we do not properly appreciate the existence of atypical forms.

Amaurotic family idiocy usually attacks children in the second half of the first year. It is characterized by increasing flaccidity of the muscles, idiocy, and a peculiar change in the eyes; is associated with convulsions and spasms, and terminates fatally within a few years. Since Sachs described the disease for the first time, in 1898, about ten cases have been reported, some of which have been studied post mortem. Tay had previously called attention to certain symptoms of the disease, which at Higier's suggestion, received the name of Tay-Sachs disease.

Amaurotic family idiocy is usually observed in several children of the same family, although a number of isolated cases have been described. Of 64 cases collected by Falkenheim, 27 were isolated and the remaining 37 occurred in members of 13 different families. It is a remarkable fact that the disease occurs with preponderating frequency among Jewish children, particularly among the children of the poor Polish and Russian Jews. The preponderance of the Jewish element is clearly shown by the fact that Falkenheim found only four out of 36 families that were Christians. Another remarkable fact is that the disease has been most frequently observed among poor immigrants in North America, while comparatively few observations have been made in the countries from which these people emigrated (so far not a single case has been reported from Vienna).

The characteristic symptoms are as follows: In a normally developed child arrest or impairment of the motor function is noticed. The child is unable to raise its head, sitting up becomes impossible, there develop flaccidity and loss of motion in the extremities and as a consequence the child is no longer able to walk or stand. While these symptoms may be attributed to rachitis, a change in the mental faculties which develops at the same time or soon afterwards attracts attention. The child forgets how to laugh, it no longer speaks to its parents and acquaintances, and takes no interest in its toys. Intellectual impairment increases rapidly. Apathy persists and is interrupted only by such occasional causes as hunger or the evacuation of urine and feces, and the child even finds some difficulty in nursing from the bottle, although the sense of taste usually remains intact even in advanced stages of the disease (the child refuses to take unsweetened milk). Finally the picture of severe complete idiocy develops.

The rapid mental decay is the more noticeable to the parents because indications of failing vision manifest themselves at the same time or soon afterwards. Ophthalmoscopic examination reveals a very peculiar, sharply circumscribed, pale white discoloration of the macula lutea, the fovea centralis persisting as a cherry-red point in the centre of the blind spot. This condition is absolutely peculiar to amaurotic family idiocy. True atrophy of the optic nerve is also noted and is rarely preceded by neuritis. Among other ocular symptoms present are strabismus and nystagmus. A symptom that is quite common is abnormal sensitiveness to noises. As the disease progresses the flaccid muscular paralysis, which is not accompanied by atrophy, is replaced by spasms with exaggeration of the reflexes. Toward the end general convulsions or tonic muscular spasms may develop. Death ensues after increasing general emaciation from marasmus or some intercurrent disease (pneumonia). The duration of the disease varies between eighteen months and two years, and the patients do not, as a rule, live to be more than two or three years old. Remissions are rare and have no influence on the ultimate outcome.

The pathology of amaurotic family idiocy consists in wide-spread degeneration of the ganglion cells and fibres of the central nervous system. Inflammatory changes do not occur. The distribution of the degenerative process is not always the same; in some cases the cerebral cortex, in others the medulla oblongata or spinal cord are chiefly affected. Sachs' original theory of a congenital disturbance in the central nervous system, as well as the theory of acquired inflammatory changes, has now been abandoned by most authors who have studied the subject. The disease represents a typical abiotic process, in which the primary factor is the change in the ganglion cells, while the degeneration of the nerve fibres is a secondary condition. In the same way, the ocular changes are regarded as a degeneration of the retinal cells; the fovea centralis, which is devoid of cells, remains intact. With good reason, Schaffer explains amaurotic family idiocy by the theory of Edinger that the tissues of the normally or functionally weak nervous system are rapidly used up and insufficiently replaced, a theory which renders this mysterious disease somewhat more intelligible.

The diagnosis is not difficult if the possibility of the disease is borne in mind. The changes in the macula are absolutely positive, and all cases reported without the characteristic ocular findings are doubtful and have in part been already acknowledged as diagnostic errors.

The disease is hopeless and no known treatment is of any avail. Fortunately it does not attack all the children in the unhappy families in which it makes its appearance.

Amaurotic idiocy presents certain noticeable points of similarity to infantile spinal muscular atrophy. Both diseases occur in very young children; in both the course is rapidly progressive; in both the termination is absolutely fatal; both present sharply defined clinical pictures such as are seen in no other endogenous disease. If amaurotic idiocy is a typical example of the abiotic processes in which the defect of the nervous system prevents the proper renewal of nervous material used up in carrying on the functions of the body, as Edinger assumes, the same thing may be true of infantile muscular atrophy. In the former, the morbid process affects chiefly the cells of the cerebellum, in the latter exclusively those of the spinal cord. From other endogenous diseases both affections are distinguished by rapid exhaustion of the resisting power of the nerve elements.

In this connection mention may be made of the occurrence of family or hereditary diseases of the optic nerve (Lebert, Higier, etc.). In these conditions inflammation and atrophy of the optic nerve occur and may lead to complete blindness. Other changes of the central nervous system are not observed in this disease, which is sometimes associated with deformities of the skull (steeple skull). The disturbance occurs in older children and in adolescents.

(c) *Periodic Paralysis of the Extremities* (*Myoplegia*)

This curious disease was first described by Hartwig in 1874 and later studied by Westphal, Oppenheim, Goldflam, Taylor, Mitchell, Oddo and Audibert, Singer, Schlesinger, Buzzard, Fuchs and Infeld. It is characterized by the paroxysmal occurrence of flaccid paralysis or weakness of the entire muscular system, involving speech and deglutition; weakness of the abdominal muscles and interference with defecation and urination. The reflexes are abolished; the electric irritability is diminished. Rarely transient spastic phenomena are present. The heart also is sometimes attacked, while, on the other hand, the muscles of the eye, face and diaphragm usually escape. The attack lasts from a few hours to three days, and the intervals of freedom are measured usually by days or weeks, rarely by months. The attack is often accompanied by sweating. Unusual muscular inactivity or overexertion appear to favor the occurrence of an attack; thus many of the patients have their attacks after certain days in the week on which they deviate from their usual mode of life. Buzzard and Singer have described cases in children. The intensity of the attacks is variable and usually diminishes as time goes on. No autopsies have as yet been performed on patients of this kind and the pathology of the condition is still in doubt. Most authorities incline to the theory of an individual predisposition to autointoxication, evidenced by occasional finding of acetone and renal elements in the urine, the occurrence of sweating and the favorable influence on the disease said to be effected by stimulating diuresis during the intervals of freedom.

(d) *Myotonia Congenita* (*Thomsen's disease*)

In 1876 the German physician, Thomsen, described a disease which had attacked several members of his family and which is characterized by muscular rigidity coming on at the beginning of an intentional movement. Since then our knowledge of this affection has been increased and confirmed by contributions from numerous authors, Seeligmüller, Strümpell, Westphal, Möbius, Bernhardt, Eulenburg, Schiefferdecker, Schultze and especially Erb. The disease is in most cases congenital. Even in those cases in which symptoms develop late there is probably a congenital predisposition, causing the appearance of morbid symptoms after severe bodily exertion. The disease is rarely recognized in early childhood (Friis) and usually manifests itself when the boy begins to receive instruction in gymnastics, at the time of military service or some unusual bodily exertion. Hence it is the fate of these unfortunate sufferers of myotonia to be taken at first for maligners.

Hereditary predisposition is usually very marked. Men are attacked more frequently than women. The tendency of the disease to occur in different members of the same family is shown in the accompanying diagram taken from Erb. The preponderance of the male sex, however, does not appear from the diagram.

FIG. 39.

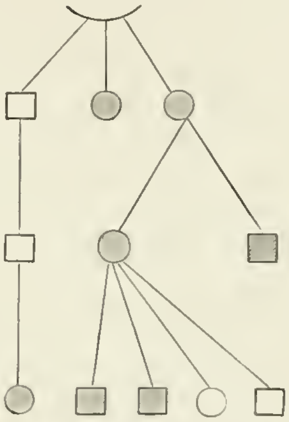


Diagram showing the influence of heredity in the case of a family with Thomsen's disease (after Erb) ○ female; ☐ male. The shaded circles and squares indicate diseased individuals.

When the disease is fully developed, any rapid movement, setting in motion muscles that have been at rest, is accompanied by a sudden stiffness, which forces the patient to persist in the spastic position. When the patient shakes hands he is unable to let go; when he suddenly takes a step forward the feet remain rooted to the ground; when he opens his mouth to masticate his food, the jaws are arrested in a half-open position. Even in beginning to speak and read the sluggishness of the muscles of the tongue and eye becomes apparent. After an interval, varying from several seconds to a half minute during which the muscles remain rigid and the patient makes awkward movements to overcome the rigidity, the muscles relax and the desired movement can be executed without further trouble; even such sustained muscular movements as dancing can be performed without difficulty. Cold, excitement, and the sense of being observed appear to favor the occurence of the muscular tension. Passive movements are not, as a rule interferred with. The muscles themselves are usually well developed and the patient may even be quite athletic. When the muscles are examined by percussion, a peculiar state of affairs is discovered. The tapping brings out a localized swelling, sometimes with a central depression, which persists a few seconds and then gradually disappears (Erb's myotonic reaction). Mild faradic irritation induces clonic spasm of the muscles which persists for some time after the current is turned off. Galvanic irritation is followed by sluggish contraction and sometimes the contraction is reversed (reaction of degeneration). Strong currents may induce undulation of the muscles. There are no other symptoms. Association of the disease with psychic affections, epilepsy, and muscular atrophy has been observed.

This very characteristic clinical picture is observed in children and even in infants at the breast. In the latter Friis, whose attention was called to the disease in infants by its presence in other members of the family, observed frequent, sighing respiration, inability to open the eyes, immobility of the face when the child began to cry, and interference with the movements of the legs and fingers. Inability to nurse at the breast is also mentioned as an early symptom of the disease. Friis' case did not show the myotonic reaction, although the muscles of the legs were hypertrophic.

The **prognosis** of myotonia is unfavorable as regards recovery, although periods of remission and improvement may be observed, especially in advanced age. The disease causes great disability in the choice of an occupation, and the subjects of myotonia are more exposed to accident because of their inability to make a sudden movement to save themselves at the right time. Examination of excised pieces of muscle shows a somewhat doubtful hypertrophy of the muscle fibrils and the presence of granules in the sarcoplasm (Schiefferdecker). In one case that was examined post mortem no pathologic changes were found in the nervous system. The **treatment** is hopeless. Internal medication, particularly with organotherapeutic products, has so far proved of no avail. Gymnastic exercises, massage, and warm baths have been employed with doubtful success.

TREATMENT OF THE ENDOGENOUS DISEASES

The treatment of the endogenous diseases is not very hopeful. The inability to give any prophylactic advice is actually depressing, and the physician has to look on hopelessly and see one child after another attacked by a family paralysis. Even Edinger's exhaustive theory has but a limited application in endogenous diseases, since it appears that even a minimum of function is enough to exhaust the imperfectly developed portions of the central nervous system. In infantile spinal muscular atrophy and in amaurotic idiocy treatment of every kind is absolutely useless. In other family affections associated with spasms and atrophy advantage may be taken of Edinger's hypothesis to the extent of guarding the child, as well as its brothers and sisters that have not yet been attacked by the disease, against any bodily overexertion, particularly by forbidding all forms of athletic exercise, including gymnastics, mountain climbing and swimming. It is better to give up the idea of keeping the child "in training" by constant use of the already damaged muscles, and to insist at once on the use of a wheel-chair or other vehicle for any extended walk. Even though the prospect of a cure is hopeless, the psychic treatment of the children should not be neglected. We should keep up the hope of ultimate recovery and not allow them to suffer from the loss of proper schooling nor cut them off from intercourse with other children.

Electric treatment in many cases has but a psychic effect. In slowly progressing muscular atrophy, however, it is not altogether hopeless and may perhaps bring about remission or even local improvement. The labile cathode or the faradic current are employed on the paralyzed extremity and the spine is subjected to stabile galvanization. In impending paralysis of the muscles of deglutition an attempt should be made, by conducting an interrupted galvanic current through the throat, to induce movements of deglutition artifically. In myasthenia electric treatment should not be used.

In muscular atrophy, massage ând gymnastic exercise are to be recommended; greatest stress being laid on passive movements. Fränkel's exercises for the treatment of ataxia should be tried in hereditary ataxia in the same way as in tabes dorsalis.

Warm baths are of great benefit, especially in spastic palsies, and may be combined with light massage. It is a well known fact that spastic extremities can be more readily moved in warm water than in bed. This phenomenon is attributed by Leyden to the buoyancy of water and consequent diminution of the action of gravity. Hot baths are to be avoided.

The question of sending these patients to watering places depends on the rapidity with which the paralysis progresses, as it is unwise to run the risk of their becoming helpless and unable to move while away from home. Any locality with plenty of woods may be selected; or we may advise indifferent thermal and saline waters, possibly containing a small amount of carbon dioxide. Among the thermal waters may be mentioned: Johannesbad, Neuhaus, Römerbad, Tüffer, Teplitz, Vöslau (Austria), Schlangenbad, Wildbad, Warmbrunn (Germany) Ragaz, (Switzerland); the temperature of the spring at Gastein is somewhat too high. For saline baths consult the chapter on scrofulosis. For carbonated baths Nauheim, Oeynhausen, Franzensbad and Soden may be considered.

Hydrotherapeutic measures with cold water are called for in these diseases only in the presence of other indications such as insomnia, excitability, depression and the like.

In comparison with these physiologic methods of treatment, internal medication is of very minor importance in the endogenous diseases. Experiments with organotherapeutic products such as muscle substance, thyroid gland and thymus have so far proved unsuccessful. It is needless to say that, when there is even a faint hope of a syphilitic etiology, the iodides and bichloride of mercury must be prescribed. As the disease is usually painless in the beginning, there is no occasion for any further symptomatic treatment. Of course, after the patient has been bedridden for some time, a number of therapeutic measures are called for, which, however, do not belong directly to the subject of neurology.

SECTION III.

HEREDITARY SYPHILIS OF THE NERVOUS SYSTEM

Hereditary syphilis may attack any portion of the central nervous system. The morbid symptoms may develop in the fœtus, and in the early years of life, in late childhood and in adolescence. It is needless to say that intra-uterine changes of the central nervous system assume practical importance only when they do not exclude the viability of the fœtus; accordingly, the meningitic, encephalitic and myelitic changes which are observed in still-born and macerated fœtuses in the brain and particularly in the spinal cord (Gasne, Gangitano and a few unpublished reports) may be dismissed with a brief mention. It is possible that early syphilitic changes in the blood vessels may lead to imperfect development of the brain and to malformations (Ilberg). A greater interest attaches to the varied morbid phenomena which make their appearance in the first years of life and for which we have at our disposal an abundance of pathological material. On the other hand, it is a noteworthy fact that in the first half year of life, when the various manifestations of hereditary syphilis are most marked, disease of the central nervous system is quite rare; analogous to the well-known fact that organic changes of the nervous system are usually absent in the secondary stage of syphilis in adults. From the 10th to the 14th year inherited syphilis begins to manifest itself in a variety of clinical conditions, some of which are quite characteristic and the accurate study of which during childhood—I need only mention tabes and progressive paralysis—is a product of recent years. The recognition of these manifestations of Fournier's late hereditary syphilis may present considerable difficulty when the patient is no longer a child. Thus, nervous affections of a hereditary syphilitic character have been observed in individuals in the third decade of life.

Diseases of the nervous system are not common in hereditary syphilis. Heubner gives them the seventh place in order of frequency among the organic diseases due to hereditary syphilis, affections of the heart and blood vessels, many of which are closely related to the nervous affections, occupying the sixth place. According to Rumpf nervous symptoms appear in about 13 per cent. of all children affected with hereditary syphilis.

Pathology.—Hereditary syphilis gives rise to a variety of changes in the nervous system, depending on the seat of the lesion and the nature of the pathologic processes that have produced the injury. In the main, the pathologic findings may be subdivided into the following groups:

1. *Syphilitic endarteritis* (Heubner) occurs both in the vessels at the base of the brain and in the small vessels of the brain and spinal cord. The vessels appear thickened; the lumen is diminished, the inner coat shows deposits and the perivascular connective tissue is hypertrophic; sometimes whole segments of some of the larger vessels may be completely obliterated (Chiari, Dowse, Kohts, Heubner).

2. *Disease of the outer and inner coverings of the central nervous system* is a peculiarity of hereditary syphilis. Inflammation of the meninges (sometimes of a hæmorrhagic character) in the form of diffuse and basal meningitis (Siemerling, Böttger, Jürgens, Bechterew, etc.), or as a chronic localized process with gummatous infiltration and thickening of the meninges (especially the pia) is found not infrequently both in the brain and in the spinal cord. In the same way the ependyma of the ventricles and the covering of the chorioid plexus may be attacked by a chronic granulating inflammation (Sandoz), which produces an accumulation of fluid within the ventricles in the form of hydrocephalus.

3. *Encephalomalacia, encephalitis,* particularly of the cerebral hemispheres, and *myelitis* occur as the result either of the endarteritis or of the inflammation of the meninges, and may lead to the production of adhesion between the meninges and the nerve substance (meningo-encephalitis), vascular neoplasms, proliferation of neuroglar tissue or degeneration of nerve fibres and cells.

4. Isolated *gummata* occur both in the cerebrum (Cnopf) and in the spinal cord, but are not frequent. On the other hand, gummatous infiltration of the meninges, of the nervous tissues and of the cranial nerves is not rare.

5. In syphilitic affections of long standing, circumscribed or diffuse *sclerotic processes* often develop in the central nervous system. The diseased portions of the cerebrum become firmer than normal, the convolutions diminish in size and may undergo pronounced atrophy (Bechterew, Busz, Shukowski, Bullen, Jacobson, Moncorvo and others).

6. *Apoplectic cerebral hæmorrhage* is rare (Nonne).

7. In the *peripheral nervous system,* affections due to hereditary syphilis are relatively rare; the nerves at the base of the brain may exhibit gummatous infiltration, degeneration and atrophy. Multiple neuritis has never been positively demonstrated post mortem in hereditary syphilis.

8. Finally, it is believed that parental syphilis may produce universal degeneration of the central nervous system without any well defined pathological findings, a degeneration which manifests itself in a variety of diseases known as *parasyphilitic affections.*

These pathological changes in the central nervous system are usually combined in a variety of ways—a peculiarity of diseases of the central nervous system due to hereditary syphilis. Clinically this same tendency manifests itself in the production of a great variety of clinical pictures, second only in that respect to the endogenous diseases. Affections of the central nervous system due to hereditary syphilis often extend over many years and their clinical manifestations during this time may be subject to great change.

Accordingly no attempt will be made in the following presentation to give a complete picture of hereditary syphilis in the central nervous system, and only the most frequent symptoms and most important clinical conditions will be discussed.

SYMPTOMS OF SYPHILIS OF THE CENTRAL NERVOUS SYSTEM

1. *Convulsions, Epilepsy.*—Convulsions are frequent as concomitant symptoms in cerebral processes due to hereditary syphilis. They may usher in the cerebral disease, they may occur as secondary symptoms in severe affections involving the brain, or represent the terminal stage in a protracted illness, and quite often the direct cause of death in children who are the subjects of hereditary syphilis. Occasionally they begin as unilateral convulsions. Cortical or Jacksonian epilepsy is quite characteristic of syphilitic diseases of the brain. Sometimes the motor spasm is preceded by sensory phenomena, formication or the sensation as if the part were going to sleep; in rare cases the attack is limited to manifestations of this kind (sensory Jacksonian epilepsy). The affected extremities sometimes become temporarily or permanently paralyzed. Attacks of cortical epilepsy may go on to general convulsions. Cortical epilepsy is due to disease of circumscribed portions of the cortex or of the meninges, such as often develops in hereditary syphilis; if the disease progresses and additional portions of the brain become involved, other cerebral symptoms are superadded. Jacksonian epilepsy therefore usually represents merely one stage in the course of some severe cerebral lesion (Soltmann, Kowalewsky, Fischl, etc.). Recurring convulsions like those which characterize genuine epilepsy not infrequently occur in hereditary syphilis. In an institution for the treatment of epileptics Bratz found that 5 per cent. of the 400 young inmates were the subjects of hereditary syphilis. Although, as a rule, epilepsy due to hereditary syphilis is accompanied by other cerebral symptoms such as disturbances of speech, tremors, rapidly progressing idiocy; apparently uncomplicated cases of epilepsy have also been observed in which a correct diagnosis was arrived at only as the result of treatment with potassium iodide (Fournier). Many otherwise typical cases of geniune epilepsy are regarded as parasyphilitic affections.

2. *Headache* is quite common, particularly in the later stages of syphilis. It may be the only symptom observed for a long time and sometimes assumes the character of hemicrania (Fournier, v. Halban).

3. *Imbecility* going on to complete idiocy is quite common in children who are the subjects of hereditary syphilis. It may be congenital or develop gradually. While in rare cases idiocy may be the only symptom ("parasyphilitic" disease), it is more frequently associated with other syphilitic symptoms, such as pupillary rigidity, convulsions and palsies, presenting either the picture of progressive paralysis (see below) or of a complicated brain syphilis. In children a disease which is accompanied by acquired progressive dementia should always arouse suspicion of syphilis.

4. *Pupillary rigidity.*—Loss of pupillary reaction and particularly loss of reaction to light while that to convergence is preserved (Argyll-Robertson pupil) is very characteristic of all forms of syphilitic processes and, barring rare cases of brain tumor and total blindness, practically always justifies the diagnosis of syphilis. In doubtful cerebral processes occurring in childhood this symptom decides in favor of syphilis. A subject of hereditary syphilis may for years present pupillary rigidity as the only symptom of the prodromal stage of tabes or paralysis.

5. *Ocular palsies*—rarely of single muscles and then only temporarily—occur in the course of cerebral diseases due to hereditary syphilis either as focal or as basal symptoms. Optic neuritis, choked disc, atrophy of the optic nerve and a chorioretinitis which is characteristic of syphilis also occur. Strabismus and nystagmus are frequently observed.

6. *Palsies of other cranial nerves,* especially of the facial, are also quite common.

7. *Speech disturbances* of every variety, such as dysarthria, bradylalia, aphasia and as symptoms of diminished mentality are quite frequently observed in hereditary syphilis of the central nervous system.

8. In the *extremities* children suffering from hereditary syphilis exhibit a great variety of symptoms. Palsies of a spastic character occur in the form of hemiplegia, paraplegia and diplegia. A monoplegia of central origin, particularly when involving the arm, is suspicious of syphilis (and brain tumor). Athetosis, tremor and ataxia are often associated with the paralysis.

9. *Sensory disturbances* are rare, and if the intelligence is preserved, the sphincters of the bladder and rectum usually escape.

10. The deep *reflexes* in the extremities are frequently exaggerated. Occasionally (tabes) they may be diminished.

The above symptoms have a tendency to combine in various ways and to form more or less sharply circumscribed clinical pictures from which it is sometimes possible to infer the existence of certain definite pathologic changes.

A. *Meningitic Affections.*—Hereditary syphilis may produce the picture of acute and chronic meningitis. Acute meningitis in rare cases develops with symptoms similar to those observed in tuberculous meningitis; in fact it has happened that a case diagnosed as tuberculous meningitis and given up by several physicians has recovered under treatment with potassium iodide given as a last resort, the effect of treatment affording the first indication of the true character of the disease (Fournier, personal observation). Most probably we have to deal in such cases with an uncomplicated serous meningitis, which has been occasionally demonstrated post mortem as an accompaniment of syphilitic changes in the brain (Caro, Gowers). Chronic meningitis of the base of the brain produces rigidity of the neck (opisthotonos in the meningitis of the posterior fossa of the skull), ocular palsies, etc. Localized meningitis of the convex surfaces of the cerebrum, which is almost always associated with encephalitis, leads to Jacksonian epilepsy and paralysis. Hæmorrhagic pachymeningitis may run a course similar to that of hydrocephalus.

B. Hydrocephalus.—That simple uncomplicated hydrocephalus may develop after birth on a foundation of hereditary syphilis is an indubitable fact (Elsner, Heller, Fruinsholz, Hochsinger, and many others). On the other hand, the relation existing between congenital hydrocephalus and hereditary syphilis is less clear. It has been attributed to intra-uterine disease of the blood vessels or to a parasyphilitic predisposition, the latter supposition finding support in the association of congenital hydrocephalus with other malformations in children suffering from hereditary syphilis (Katzenstein) (see congenital hydrocephalus). Acquired hydrocephalus when due to hereditary syphilis usually makes its appearance in the first year, particularly during the first six months of the infant's life. The symptoms are extremely variable. In many cases, the only symptoms are enlargement of the skull and bulging of the fontanelle. The intelligence is not necessarily impaired. In other cases hydrocephalus begins like a meningitis with symptoms of brain irritation such as restlessness, insomnia, vomiting, rigidity of the neck and convulsions, and quite frequently leaves a permanent rigidity with contractures of the extremities and marked exaggeration of the reflexes. As a rule, there is imbecility or idiocy. Enlargement of the skull in all these cases is very marked and may attain extreme degrees (56.5 cm. Hochsinger); but on the whole acquired syphilitic hydrocephalus does not produce an extreme dilatation of the skull so frequently as the congenital form. When the enlargement is very great, the child is unable to raise the head. The eyes often exhibit the peculiar positions already described in connection with congenital hydrocephalus. The remaining symptoms that have been described in connection with congenital hydrocephalus may also be present in the acquired form.

The course of syphilitic hydrocephalus is subject to many variations. In rare cases the meningeal symptoms undergo acute exacerbation and the disease terminates suddenly in death (Fruinsholz); or the course may be subacute, extending over several months, and ultimately ending in death (d'Astros). Now that the etiologic relation between hereditary syphilis and hydrocephalus has become better known more cases are cured, or at least arrested by antisyphilitic treatment (cases of Hochsinger, Heller, Neumann, Immerwol and others). Unfortunately the cure is not always permanent, and a fresh attack of hydrocephalus or other signs of brain syphilis often develop after an interval of several months. Finally, hydrocephalus in many cases causes a permanent condition characterized by general convulsions and imbecility which leaves the children in ill health or at least very much underdeveloped.

C. Cerebral Infantile Palsy.—The various clinical pictures of cerebral infantile palsy very frequently rest on a foundation of hereditary syphilis. I have personally known cases of this kind presenting hemiplegia, paraplegic rigidity and athetosis and König, who was at first inclined to doubt the influence of hereditary syphilis in the production of cerebral infantile palsy was able to demonstrate its presence in seven per cent. of his cases. General rigidity, noted soon after birth and attributed to injuries during labor, is also observed in children affected with hereditary syphilis (Rolly, Vizioli, Gilles de la Tourette and others). These palsies are usually associated with idiocy, epilepsy, and sometimes pupillary rigidity. These forms of cerebral infantile palsy, which rest on a foundation of hereditary syphilis, should be differentiated from cerebral infantile palsy proper because, as we shall see, the latter is equivalent to a healing process within the brain, whereas those forms of cerebral palsy which rest on a syphilitic basis may at any time undergo a change, and usually a change for the worse.

D. Brain Tumor.—The symptoms of brain tumor are sometimes produced by hereditary syphilis in the form of gumma or encephalomalacia (Cnopf; case of my own in a syphilitic boy presenting distinct symptoms of a pontine tumor). Antisyphilitic treatment is not always followed by favorable results; both idiocy and death have been observed.

E. Multiple Sclerosis.—The familiar symptom-complex of multiple sclerosis—scanning speech, intention tremor, nystagmus, disturbance of gait and mental deficiency—although not characteristic in childhood, is not infrequently due to hereditary syphilis. In such cases we may assume a multiple focal encephalomyelitis or a diffuse sclerosis (Bechterew).

F. Tabes Dorsalis and Progressive Paralysis.—By including tabes and paralysis in the chapter on diseases of the central nervous system due to hereditary syphilis we wish to emphasize the fact that, so far as the pediatrist is concerned, there is practically no doubt of the etiologic relationship between syphilis and this condition. This is clearly proven by statistics. Marburg was able to demonstrate the presence of syphilis in 26 out of 34 juvenile tabetics; Alzheimer found positive, or very suggestive signs of syphilis in 70 per cent. of 41 juvenile paralytics, and these figures assume greater significance from the fact that Hirschl, on questioning patients with tertiary syphilis in a syphilitic ward, obtained a history of a former attack of syphilis in only 54 per cent. A still more trenchant argument is found in the fact that progressive paralysis and tabes have never been observed in any child in which syphilis could be positively excluded after careful observation from the first day of life. From this standpoint the frequent association in childhood of two diseases which in themselves are not frequent—hereditary syphilis on the one hand and tabes or paralysis on the other—must be regarded as something more than a coincidence and the cases, by no means rare, in which the first signs of hereditary syphilis and the initial symptoms of tabes or paralysis have been demonstrated in the same individual when under continuous observation (Hochsinger) must be regarded as positive proof of a causal relationship which is indubitably constant. At all events the conditions found in childhood are of considerable importance in helping to decide this question in adults (K. Mendel).

When it is remembered that in the adult from 15 to 20 years usually elapse between the syphilitic infection and the first signs of the above-mentioned nervous affections, there is nothing surprising in the fact that in the child these diseases do not begin to develop until the end of childhood, so that the first obvious signs are often observed during puberty. On the other hand, there is a tendency to a shortening of the interval in children, and paralysis especially is often seen fully developed in children of ten to twelve years of age. The incidence of tabes or paralysis is the same in boys and girls (Marburg gives the proportion of girls to boys as 4:3; Alzheimer for paralysis as 1:1). This again is in accord with the theory of heredity, whereas in adults, owing to the greater frequency of syphilitic infection in men, the number of male tabetics and paralytics is greater then the number of females afflicted with these diseases.

Tabes and paralysis present characteristic forms in childhood and usually occur separately. There are cases, however, which, as in the adult, present common symptoms of the two diseases and which may be designated tabo-paralysis. Many cases that would now be regarded as cases of paralysis or tabes were formerly described as forms of cerebrospinal syphilis, which somewhat complicates the task of sifting the material.

(*a*) *Tabes Dorsalis.*—The number of cases of this disease so far described is not large (Marburg has collected 34 cases). This is due partly to the comparative rarity of the affection and partly to the difficulty of making the diagnosis, chiefly because the disturbance of the gait, which is such a prominent symptom in adults, is rare in the juvenile form. Among the cardinal symptoms in tabes failure of the pupil to react to light is very frequent, there being no difference in that respect between the infantile and the adult forms. Inequality of the pupils is common. The patellar reflex is usually abolished, but this symptom is not as constant as in adults. Swaying when the eyes are closed is rarer in juvenile tabes than in the adult form. Ataxia and disturbances of the gait are much less marked and are observed only in a small minority of the cases of infantile tabes. On the other hand, genuine atrophy of the optic nerve, incontinence of urine—first in the form of nocturnal enuresis—patches of anæsthesia on the trunk and lancinating pains in the legs are not rare in children. Gastric crises, neuralgias, hemicrania and trophic changes (arthropathies, mal perforant) have also been observed both as prodromal phenomena and as symptoms of the disease when fully established.

The onset of infantile tabes, as may be inferred from the symptoms described, is even more indefinite than in adults. The patients are hardly inconvenienced by the initial symptoms; lancinating pains are interpreted as rheumatism, atrophy of the optic nerve as beginning myopia and the incontinence of urine as enuresis. Only when some more striking symptom such as headache or increasing visual disturbances develops, a physician is consulted, and the history and physical examination reveal a tabes of long standing. The course is extremely tedious and may extend over several decades. As the first visible signs usually develop during the latter part of childhood (in exceptional cases between the fifth and sixth years, Dydyski, Idelsohn and v. Rad), tabes due to hereditary syphilis usually runs its manifest course after the age of childhood. So far as I know, there are as yet no post-mortem reports of juvenile tabes (quite recently Köster described a case with changes in the posterior columns). Treatment with the iodides and bichloride of mercury is followed by no better results than in the adult form.

(*b*) *Progressive Paralysis.*—Our knowledge of progressive paralysis in childhood being of less recent date than our knowledge of tabes, the number of cases is correspondingly greater. In 1898 Thiry collected 69 cases, and since then a considerable number of additional cases have been contributed to the literature. I myself have had the opportunity of seeing from 6 to 8 cases of this disease.

Progressive paralysis usually begins between the 12th and 16th years of life, the lowest limit being about 8 (or, according to Zappert, five years), the upper limit twenty years. Patients in whom the disease makes its appearance toward the end of childhood or even later very frequently exhibit an infantile habit and delayed puberty, as is so often observed in cases of hereditary syphilis.

The symptoms of the disease are partly psychic and partly somatic. Dementia, which is of sudden onset, occupies the most prominent place in the picture. In contradistinction to progressive paralysis in adults, the mental disturbance in children manifests itself in steadily increasing mental decay without fixed ideas or hallucinations. The children become apathetic, lose interest in their surroundings and in their toys, become undemonstrative and give no sign of affection for their parents, forget all they had learned either in school or at a simple trade, are sluggish in their movements and, when left to themselves, idle their time away aimlessly, taking up and looking at any object that comes in their reach without betraying any knowledge of its proper use. They are subject to occasional outbreaks of passion, crying fits, uncontrollable rage, with or without adequate reason; but these attacks never degenerate into mania. Such children are never seen to indulge in hearty laughter.

These psychic changes are accompanied by disturbances of speech. Letters and syllables are confused; there is bradylalia, babbling speech (lallatio); the child's vocabulary rapidly diminishes, and finally it ceases to talk altogether and its utterances are limited to a few almost unintelligible words.

Along with these marked psychic disturbances certain somatic symptoms are observed. The face becomes expressionless and the child develops a tendency to indulge in sucking and chewing movements, humming and puckering the lips. Tremor of the lips and tongue is often observed. Pupillary rigidity is usually present early in the disease. The patellar reflexes are almost never absent and frequently exaggerated; paralysis of the extremities is never observed, at least during the initial period of the disease.

A special peculiarity of infantile paralysis is the early occurrence of paralytic attacks. These manifest themselves in the sudden onset of vertigo and loss of the power of speech, temporary unconsciousness, paroxysmal headache and epileptiform seizures. In rare cases the paralytic attack is preceded by hemicrania, paralysis of the extremities and ptosis. All the above-described symptoms may be regarded as the initial indications of progressive paralysis and sometimes persist from 1 to 2 years without exhibiting any change or, at most, a slow progressiveness. Gradually the somatic symptoms become more pronounced. The patients exhibit tremor and intention tremor and contractions of the extremities; they lose the power of walking, become bedridden and rapidly emaciate. The terminal stage is often marked by bedsores and consequent fever. The patient finally dies of general sepsis, pneumonia, or in a paralytic attack. Remissions such as are so frequently observed in the paralysis of adults are very rare in the juvenile form. The duration of the disease according to Hirschl and Thiry is 3¾ years, according to Alzheimer 4½ years, or in other words longer than in adults, in whom the average duration is two years.

Tabetic symptoms, especially atrophy of the optic nerve, absence of the patellar reflex and lancinating pains, may be superadded to those of progressive paralysis. According to Alzheimer cases of this kind have a longer duration than cases of simple paralysis.

Numerous observations have been made on the **pathology** of progressive paralysis (Alzheimer, Hirschl, Meyer). The brain is atrophic, the convolutions are narrowed, the pia greatly thickened, cloudy and adherent to the cortex; the basal ganglia often exhibit marked atrophy. Microscopically we find pronounced leptomeningitis, syphilitic endarteritis, increase of connective tissue in the meninges and in the brain, and considerable atrophy of nerve fibres, particularly in the deeper layers of the cortex; the radiation of fibres in the medulla is diminished. In the spinal cord sclerotic changes are sometimes seen in some of the columns.

Treatment is absolutely powerless in progressive paralysis.

G. Other Diseases of the Spinal Cord due to Hereditary Syphilis.—In addition to the affections in which spinal symptoms are associated with disturbances of the cerebral function (Siemerling, Böttger, Lamy, Sachs, etc.), and in addition to tabes dorsalis, which we have been discussing, children affected with hereditary syphilis exhibit certain conditions which by their clinical course as well as by their pathology, point to some lesion of the spinal cord alone.

From the pathologic investigations of Gasne and Gangitano already referred to, it appears that newborn syphilitic children often exhibit extensive pathologic changes in the spinal cord of the nature of a diffuse or a circumscribed meningomyelitis, which is usually associated with other organic diseases and destroys the child's life. According to Gilles de la Tourette, spastic paralysis may result from this condition if the child survives and may simulate the clinical picture of universal rigidity (spastic paraplegia).

More important than the above are those cases of spinal syphilis in which clinical symptoms develop late. Gilles de la Tourette recognized two groups, depending on whether the disease begins in earliest childhood or at a later period of life. The symptomatology of these conditions consists in paraplegia of sudden or gradual onset, pain, sensory disturbances, flaccid atrophic palsies and disturbances of urination and defecation. The grouping of the symptoms may be such as to suggest, in some cases, circumscribed myelitis or gumma of the spinal cord; in other cases, a diffuse inflammation extending over the entire cross-section of the cord. Cerebral symptoms usually make their appearance at the same time.

Among special forms of spinal syphilis the following should be emphasized: Fournier described what appeared to be a spondylitis, followed by paresis of the legs which yielded to treatment with the iodides. Hoffman reported a case of simple spastic spinal paralysis resembling the spastic spinal paralysis observed in the syphilis of adults, and described by Erb (cases of von Mendel, Luzenberger, Sachs, Nonne, etc.).

Friedmann knows of cases with repeated attacks of spastic paralysis in the legs, with weakness of the bladder, lasting several months, followed by temporary recovery. He regards the condition as a peculiar *relapsing form of spastic spinal paralysis* depending upon hereditary syphilis.

The prognosis in these cases of simple spinal syphilis is not so unfavorable as that of tabes. Vigorous treatment with the iodides and bichloride of mercury is often followed by arrest or complete cure of the disease. The cases in which the disturbances are congenital or develop in earliest childhood are more unfavorable.

II. Peripheral Palsies.—*Multiple neuritis* due to hereditary syphilis is unknown. One case reported by Nonne, although quite characteristic, lacks the proof of a syphilitic basis. Palsies of individual ocular muscles of temporary character (Zappert), pupillary rigidity, and diseases of the fundus have been observed as isolated symptoms, but probably represent only separate stages in the course of brain syphilis. Deafness of rapid onset and due to central origin (labyrinthina deafness) is a well known late symptom of hereditary syphilis.

For *syphilitic pseudoparalysis* see the chapter on hereditary syphilis. There is no doubt that this temporary paralysis of the arms in infants in the majority of cases is due to syphilitic osteochrondritis; but the peculiar type of this reflex paralysis and the occasional occurrence of cases without any demonstrable bone lesion are worthy of note. Since, however, palsies of this kind are associated with contractures (Doucas, Demetriades, Reuter), with ptosis (Sandoz, Soltmann), with oculopupillary symptoms (? Peters), and in view of the absence of bone lesions as shown at the autopsies of otherwise typical cases (Scherer), it is a question (Henoch, Heubner, Reuter and Pollack) whether diseases of the bone can be properly regarded as the cause in all these cases presenting the picture of a pseudoparalysis.

The question is still under discussion. A case reported by Zappert, which was unquestionably one of hereditary syphilis with circumscribed meningitis in the cervical portion of the cord and secondary paralysis of the arms, was regarded by Hochsinger as possibly a birth palsy.

I. In addition to the above-described forms of hereditary syphilis there are a number of other conditions that may, with more or less reason, be attributed to hereditary syphilis. These include observations of symmetrical gangrene (Krisowski), diabetes insipidus (Demme), of trifacial neuralgia (Soltmann), and, finally, a statement by Hochsinger that his myotonia of the newborn shows a special predilection for the subjects of hereditary syphilis.

When we consider the enormous number of different conditions that may develop in the nervous system of children affected with hereditary syphilis, the diagnostic difficulties are at once apparent. The important point for the practical doctor is to remember the possibility of hereditary syphilis in such cases, as the prospect of a cure is so much more certain if that diagnosis can be established. Aside from the history and other signs of syphilis in the child or other members of the same family, any acquired chronic nervous condition with pupillary rigidity, imbecility, and cerebral symptoms of difficult localization should excite the suspicion of hereditary syphilis. To make an accurate scientific diagnosis in such cases is far more difficult; for it is sufficiently evident from what has been said that only certain definite pictures, such as hydrocephalus, tabes, progressive paralysis and spastic spinal paralysis are sufficiently characteristic to admit of a precise diagnosis. In very many cases all attempts at an accurate diagnosis must be given up and the practitioner will have to content himself with a general determination of the seat and character of the disease.

The same caution is necessary in giving a **prognosis.** Although it is quite true that in many conditions simulating a severe meningitis, a brain tumor or a myelitis, the demonstration of hereditary syphilis offers a ray of hope and the possibility of saving the child, it must not be forgotten that many diseases due to hereditary syphilis continue to progress steadily in spite of specific treatment, and that even recovery from one syphilitic manifestation in the nervous system does not preclude the reappearance of the constitutional disease in another form. Nevertheless, it is true, that among diseases of the nervous system of equal severity, those which are due to hereditary syphilis have by far the best prognosis.

In regard to causal **treatment** directed against the cause of the disease the reader is referred to the chapter on hereditary syphilis. With the exception of tabes, paralysis and many forms of epilepsy, the effect of antisyphilitic treatment in most of the diseases that have been mentioned is quite prompt, although not always lasting. In cases in which iodine and mercury prove ineffective, the same therapeutic principles obtain for diseases of the nervous system due to hereditary syphilis as for the same diseases when not due to syphilis.

There can be no doubt that syphilis *acquired in early life* may also produce the above-described symptoms. Observations on this point are not numerous, and better proof should be obtained than the mere statement of parents, who frequently attempt to hide the fact that they are themselves syphilitic by attributing the infection to the nurse.

SECTION IV.

TUBERCULOSIS OF THE CENTRAL NERVOUS SYSTEM

(SPONDYLITIC COMPRESSION OF THE SPINAL CORD)

The child's central nervous system is very often the seat of tuberculous disease. Aside from localized pachymeningitis in caries of the petrous portion of the temporal bone, brain abscess, sinus thrombosis of tuberculous origin, and the rare form of myelitis with granular degeneration of the cells, which can only be demonstrated post mortem, tuberculosis manifests itself chiefly in three quite frequent and characteristic forms: (1) meningitis; (2) tuberculosis of the brain and spinal cord; (3) compression of the spinal cord by caries of the vertebræ. As tuberculosis of the meninges, and brain tuberculosis in connection with other diseases of the meninges, and brain tuberculosis in connection with neoplasm of the central nervous system are discussed elsewhere, we shall confine ourselves at this place to the discussion of

COMPRESSION OF THE SPINAL CORD BY CARIES OF THE VERTEBRÆ (SPONDYLITIS)

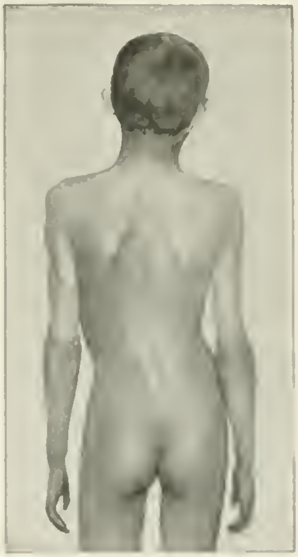
Spondylitis or Pott's disease is a frequent manifestation of tuberculosis in childhood and occurs chiefly in children with tuberculous inheritance. It is often accompanied by tuberculosis of the bones, glands, joints or lungs; but clinically, at least, it not rarely constitutes the only demonstrable tuberculous lesion present. Boys are attacked somewhat more frequently than girls; not even infants escape; the period of greatest incidence of the disease is between three and five years of age.

The disease rarely attacks only one vertebra; as a rule, 2, 3, 4 or even as many as 8 vertebræ are involved (Hugelshofer, Beuthner, Bovier and others). The most frequent seat is in the lower dorsal, or upper lumbar portion of the cord, and in small children it is quite frequently found in the cervical portion of the spinal column. Cases in which the lesion is confined to the first two cervical vertebræ have also been observed. So long as the vertebra chiefly involved retains its shape, spinal caries does not produce any demonstrable symptoms; but if the destructive process is advanced, the bone collapses under the pressure of the superimposed vertebræ and is displaced backward, producing an acute angled deformity of the vertebral column. The spine of the vertebra, which is displaced backward, becomes visible externally, forming a so-called gibbosity. The collapse of the carious vertebra may be hastened by injury, which is then erroneously regarded as the cause of the spondylitis. Carious disease of bone frequently leads to the accumulation of pus, either in the neighborhood of the diseased focus or more frequently in the form of a deep abscess in the neighboring tissues. This complication, which affects the prognosis unfavorably, is specially apt to occur in dorsal and lumbar spondylitis, being more rare in the cases in which the cervical portion of the cord is involved.

Vertebral caries assumes neurological importance only when the spinal marrow becomes involved in the morbid process. If pains produced by disease at the roots of the spinal nerves are also to be ascribed to involvement of the spinal cord, the cases will be much more numerous than if only cases with well-marked spinal symptoms are included.

The pathologic foundation of the spinal disease spondylitis is compression of the vertebræ from lack of space. The cause of this lack of space may be direct pressure of a diseased vertebra or a local abscess, but is much more frequently found in a proliferation of the granulation tissue at the carious site encroaching upon the vertebral canal or local inflammation with marked thickening of the dura. The resulting disturbance of the circulation produces œdema in the spinal marrow and finally softening. This softening of the marrow, in combination with the increasing pressure from without, ultimately produces marked contraction and sclerotic changes in the cord, so that the structure of the spinal cord may be completely destroyed at the point of angulation. Pronounced myelitis, however, occurs only in exceptional cases; hence the old designation compression-myelitis is not appropriate in the majority of cases (Schmaus). The spinal roots, especially the posterior roots, usually suffer much earlier from pressure or from the dural disease than the spinal marrow itself.

Fig. 40.

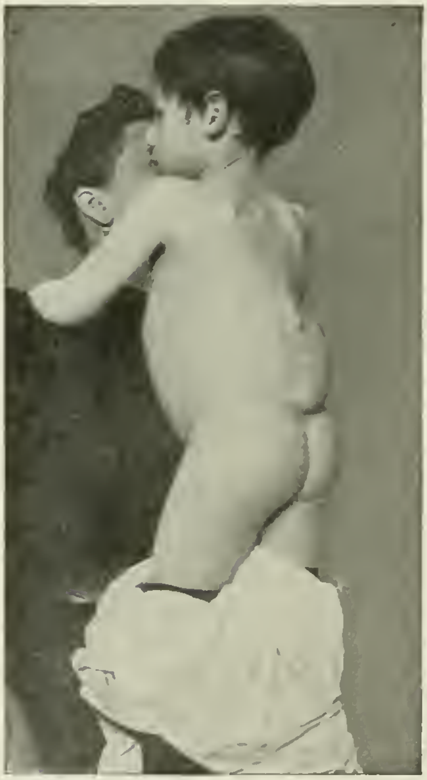


Dorsal spondylitis. Boy eleven years old. Characteristic rigidity of the vertebral column. No kyphosis.

The most frequent **symptom** of spondylitis and one which is practically never absent is pain. The pain is often the subject of bitter complaint on the part of the patient; in other cases it must be elicited by tapping the vertebral column, concussion of the thoracic cage by sudden pressure on the shoulder, or by passing a hot sponge over the spine. If there is kyphosis, the hump is usually the seat of pain, either spontaneous or elicited by pressure. Rigidity of the vertebral column is the direct consequence of the pain. If the seat of the disease is in the cervical portion of the cord, the head is rigidly held in the same attitude, sometimes inclined to one side as in torticollis; or the child supports the head with both hands and nervously resists any attempt at movement. In spondylitis of the thoracic or lumbar portion of the cord the rigidity produces a lordosis, all the movements of the body are extremely cautious, and the patient makes every effort to maintain the vertebral column in a condition of absolute rest. Direct spinal symptoms are a much more rare occurrence than pain in spondylitis (in 10.7 per cent., Hugelshofer; 12.7 per cent., Vulpius). The character and distribution of the various symptoms depend on the seat and intensity of the disease.

The most frequent **localization** of the carious portion is in the dorsal and lumbar portions of the cord, and the first symptom in this case is rigidity of the legs, exaggeration of the reflexes, with weakness and paraplegia. To these symptoms are superadded in the subsequent course of the disease, diminution of sensibility in the legs, disturbance of the bladder and rectum, bedsores, and, in short, all the symptoms with which we are familiar in the adult as the result of a transverse myelitis. Fortunately, the disease in most cases does not progress beyond a spastic paraparesis. If the lesion causes great destruction of the spinal marrow in the lumbar enlargement, atrophic paralysis of the legs with diminution of the reflexes may result. If the lesion is situated in the cervical portion of the cord, all the extremities become involved in the spastic paresis; and again, if the cord suffers direct compression, a combination of atrophic paralysis of the arms with spastic paresis of the legs may be observed. Lesions of the sphincters are usually absent; on the other hand, paresis of the thoracic or abdominal muscles is not uncommon, and the phrenic nerve and diaphragmatic action may even be threatened. In those rare cases associated with destruction of the highest cervical vertebræ, rigidity of the neck and fixation of the head movements are more pronounced than in any other lesion. In such cases there is, in addition to spastic paresis especially of the arms, paralysis of the accessory nerve and of the hypoglossus, as well as other bulbar symptoms, and sudden death may result from compression of the medulla oblongata. To sum up, the most frequent nervous complication of spondylitis, aside from the localized pain, consists in spastic

Fig. 41.



Dorsal spondylitis in a boy five years old. Well marked kyphosis with large spondylitic abscess.

paresis of the legs or, possibly, of all four extremities and, in severe cases, the picture of a complete transverse lesion of the spinal cord.

The most important **diagnostic sign** of spondylitic compression paralysis is kyphosis; if this is absent, pain referred to, or elicited in the vertebral column is, in view of the frequency of caries of the vertebræ in the child, an important diagnostic sign, provided disease of the muscles and of the peripheral nerves and internal disease can be excluded. Sometimes, particularly if pain is absent, the rigidity of the vertebral column may lead to confusion with spinal meningitis, muscular atrophy or rachitis, particularly as the peculiar manner of rising from a stooping to the erect posture, which is so characteristic of muscular atrophy, may be observed in the initial stages of spondylitis. X-ray examination very often gives quite satisfactory results, the diseased vertebræ appearing pale in the photograph, although I have occasionally been disappointed in early cases. Hysteria may have to be considered in the differential diagnosis of vertebral caries in childhood. As a rule, however, the rigidity of the back disappears on rapid movement, and the general appearance and behavior of the patient usually suggest the correct diagnosis. In doubtful cases the presence of fever, emaciation and other signs of scrofulosis and tuberculosis are of course in favor of spinal caries.

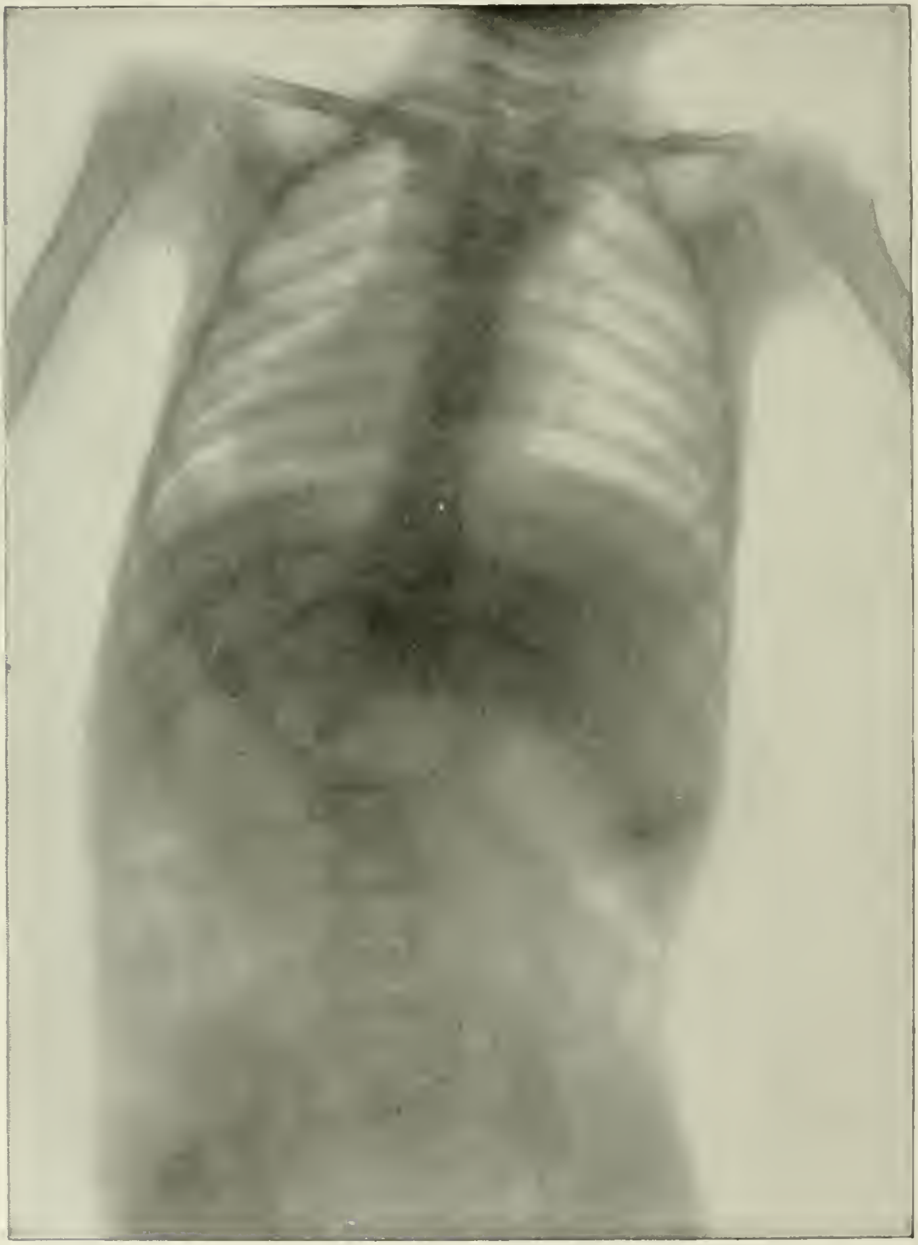
The severity of the paralysis in spondylitis is not always dependent upon the intensity of the spinal caries. Quite often very severe or even fatal tuberculous disease of the vertebræ is observed to run its course without producing any marked symptoms of paralysis.

The **prognosis** so far as the paralysis is concerned is not altogether unfavorable. In many cases a decided improvement and ultimately complete restoration of normal function occur (disappearance of œdema). When spondylitis proves fatal, death is much more rarely the result of vertebral disease (decubitus, cystitis) than of general tuberculosis such as tuberculous meningitis or amyloid disease. The mortality of spondylitis is according to Reinert 60 per cent., according to Hugelshofer 57.6 per cent. and according to Billroth 52.1 per cent.

The **treatment** of spondylitic paralysis or spinal caries has been made the subject of much study in orthopedic surgery. It is impossible to discuss the details of treatment in this place; suffice it to say, that absolute rest in bed with extension, supporting apparatus and plaster jackets are the means employed in the treatment (Glisson sling, Rauchfuss' suspensory apparatus; plaster of Paris bed after Lorenz).

Many attempts have recently been made to treat spinal caries by operative means. The most radical procedure consists in directly attacking the kyphosis (laminectomy), removing of the abscess or granular tissue compressing the spinal cord or cicatricial or thickened portions of the meninges and scraping away the carious bone. The first operation of this kind was performed by Macewen, who was so fortunate as to get good results from his first cases because they were in process of recovery. Later operators, however, satisfied themselves that in florid active tuberculous processes of the vertebræ, radical removal of the diseased tissue is frequently impossible and that, after a

Fig. 42.



X-ray photograph of a case of marked spondylitis in upper lumbar cord. Diseased portion is recognized by the pale spot at side of destroyed vertebra. The lateral inclination of the thorax is pathological.

temporary improvement in the paralysis the original state of affairs soon returns. From an analysis made by Chipault, it appears that, of 103 cases of laminectomy only 15 ended in permanent recovery—reason recent enough for extreme conservatism in deciding upon such an operation (Schlesinger). Measures directed to the gradual straightening of the prominent vertebræ are more encouraging, although their object is rather to improve the patient's appearance than to exert any influence on the paralysis which ultimately develops. Forced compression of the kyphosis, was recommended by Calot a few years ago, for a time attracted a great deal of attention, but has since been abandoned as too dangerous.

General supportive treatment such as is indicated in any form of tuberculosis must be resorted to in spondylitis also.

SECTION V.

INFLAMMATION OF THE CENTRAL NERVOUS SYSTEM

I. ENCEPHALITIS

The study of encephalitis in childhood presents some very difficult problems, some of which still await solution. The difficulty lies in the want of harmony between pathologic findings and clinical experience, in the fact that the symptoms vary not only according to the seat of the disease but also according to the patient's age and, finally, that in childhood recovery from encephalitis is probably much more frequent than in adults and it is accordingly difficult to establish a diagnosis of encephalitis from existing focal symptoms or retrospectively from autopsy changes discovered at some later period. In view of these facts we shall divide the subject loosely into the following subdivisions: *interstitial congenital encephalitis* (Virchow), *encephalitis of infants*, and *encephalitis of older children*. In discussing cerebral infantile palsy we shall have occasion to refer to infantile encephalitis again.

1. *Interstitial Congenital Encephalitis* as described by Virchow in 1865 is interesting solely from a pathologic viewpoint and its existence as a clinical entity is very doubtful. According to Virchow the characteristic feature of this form of encephalitis is the presence in the brain of fatty granule cells, which are found either diffusely or in disseminated nests in the cerebrum of newborn infants. Very soon, however, a doubt was raised (Hayem, Jastrowitz, Kramer, Flechsig and others) whether these structures were really pathologic, and it was contended that these fatty granule cells are normal in the brain of the newborn. Other authors (v. Limbeck, Fischl, Thiemich) are inclined to make a distinction between diffuse and circumscribed cellular accumulations and to accord to the latter at least a pathologic significance. From my own quite extensive investigations, which, it is true, have reference to the spinal cord rather than to the brain, I have come to the conclusion that

the fatty granule cells at a certain period of life unquestionably represent a normal condition in the central nervous system of man and accumulate in many portions of the brain and spinal cord in a manner quite remarkable. On the other hand I know from personal experience that small, yellowish foci occur in the brain of newborn infants, and since these foci, according to the investigations of several authors, contain, in addition to the fatty cells, round cell infiltration, changes in the ganglion cells and proliferating neurogliar tissue, there can be no doubt that these small foci represent an inflammatory process. It will be advisable in future histologic investigations to pay more attention to other tissue elements that are characteristic of inflammation rather than to the fatty granule cells. We may then expect a solution of the question of Virchow's encephalitis and it may be found that newborn children, particularly immature and debilitated infants, may react to general septic processes by the formation of small inflammatory foci in the brain, and that the finding of fatty granule cells is not enough to establish a diagnosis of encephalitis. At all events these findings have a purely pathologic significance, and it is very questionable whether the smaller foci ever coalesce to form large inflammatory areas capable of producing clinical symptoms; it is more probable that such conditions begin as severe and extensive lesions.

2. *Acute Encephalitis of Infants.*—Investigations by a number of authors (Gaudard, Kast, Jendrassik, Marie, Reymond, Fischl, Ganghofer, Finkelstein and others) have established the occurrence in infants of a clinical picture which begins acutely with violent cerebral symptoms, has a fatal termination and reveals post mortem an extensive inflammation of the brain.

The disease is apparently primary and occurs without any antecedent characteristic infectious disease. It is probably the result of some septic process such as are so frequent at this age. Premature, sickly children are particularly prone to diseases of this kind. The seat of inflammation is practically always in the cerebrum, particularly in the hemispheres, sometimes in the basal ganglia, and rarely in the pons.

The brain usually exhibits a severe degree of softening and often is converted into a semi-fluid, deliquescent, "creamy" or "raspberry-like" mass; or it contains numerous hæmorrhagic foci; or, finally, there may be sclerosis with narrowing of the convolutions and secondary hydrocephalus. Hence, in some cases softening, in others hyperæmia may be the chief condition, or a hæmorrhage may be simulated. Histologic examination reveals necrosis, atrophy of the white matter, disappearance of the ganglion cells, accumulations of leucocytes, masses of fatty granules, in some cases a high grade of plethora, hæmorrhage, or even a marked proliferation of neurogliar tissue.

The clinical picture is usually that of an extreme febrile symptom-complex. The temperature rises rapidly to 40° to 41° C. (104° to 105.8° F.) and continues very high. Disturbance of consciousness occurs early. There is stupor, which is soon replaced by complete coma. Convulsions are always present; they often represent the initial stage of the disease and are prone to recur again and again. The breathing is superficial. Cheyne-Stokes respiration and attacks of asphyxia occur. The pulse is usually greatly accelerated and feeble. Rigidity of the neck is frequently present; bulging of the fontanelle is not always observed. The extremities are rigid, usually in a position of flexion. Strabismus is not uncommon. These severe general symptoms overshadow the localized focal phenomena, which are usually confined to slight paresis or monospasm of individual extremities or of the facial nerve.

The duration of infantile encephalitis is from 1 to 2 weeks; the child ultimately dies of cardiac or respiratory failure.

The **differential diagnosis** between acute inflammation of the brain substance and simple acute meningitis practically cannot be made by the clinical symptoms. In the presence of a pneumonia or some intestinal process, and after pertussis the chances are in favor of meningitis. Turbidity of the cerebrospinal fluid obtained by lumbar puncture also points to inflammation of the meninges.

It is not to be inferred from the above schematic description of infantile encephalitis that acute inflammation of the brain in the early periods of life is always so extensive and necessarily runs a fatal course. It is impossible to make a sharp distinction between diffuse severe forms of encephalitis which have a special tendency to occur in early infancy and the more circumscribed conditions which belong to the later periods of life; and infantile encephalitis may run a mild course ending in recovery just as, on the other hand, the disease in older children may be marked by the above-described violent symptoms and end in death. But in order to understand the forms of encephalitis without autopsy findings which will be discussed presently, a knowledge of the fatal forms of the disease is necessary, because, with respect to the intensity and extension of the morbid process, these must be regarded as exaggerated forms of the same disease which ends in recovery.

3. *Encephalitis in Older Children with a Tendency to Recovery.*—In children between the ages of 2 and 4 acute inflammation of the brain may run a course similar to that observed in the infant. More frequently, however, it is circumscribed and the symptoms are not so violent. A large proportion of the cases terminate in recovery with persistence of focal symptoms.

The most frequent causes of encephalitis after the first and second years of life are the infectious diseases—measles, diphtheria, scarlet fever and whooping-cough. Acute meningitis is frequently followed by a superficial encephalitis. Many forms of acute inflammation of the brain which, however, are rare in childhood, are caused by intoxication. Aside from these secondary forms of diseases we also have an acute inflammation of the brain occurring primarily like an independent disease. Strümpell must be given the credit for calling attention to these conditions and his investigations have taught us to regard polioencephalitis or acute inflammation of the brain, poliomyelitis, and possibly also inflammation of the peripheral nerves as different manifestations of an independent acute disease due to bacterial causes and possessing a special predilection for childhood. The same individual not infrequently presents the remains of both cerebral and spinal palsies, and epidemics of acute inflammation of the brain and of the spinal cord in association have even been described (Medin). Head injuries may possibly be capable of producing a simple encephalitis as well as a purulent inflammation, as is shown by the occurrence of inflammation of the brain after severe injuries to the skull and also by animal experiments, although it is still doubtful whether the contusion itself is the actual cause of the inflammation or only the exciting cause acting on a previous bacterial predisposition.

There are no statistics in regard to the frequency of encephalitis in children. If we leave out other rarer causes, such as sinus thrombosis, and attribute cases of cerebral infantile palsies beginning with acute febrile brain symptoms to encephalitis, we must conclude that the disease, as well as poliomyelitis, is quite frequent in childhood and in the majority of cases ends in recovery.

For the *pathology* of these forms of encephalitis we depend in part on post-mortem findings in infants and adults, and in part on cases that have ended in recovery and have come to autopsy later, because very few post-mortem observations have been made in older children.

Acute circumscribed inflammation of the brain in its typical form is a hæmorrhagic encephalitis exhibiting one or several inflammatory foci with the characteristic histologic findings. In less recent cases this red softening is replaced by so called yellow softening, in which the foci present a necrotic appearance and contain many degenerated tissue elements, with less marked engorgement of the blood vessels. The termination of encephalitis in abscess, cysts, sclerosis and porencephaly, will form the subject of a later chapter (brain abscess, diffuse sclerosis, infantile palsy). Bacteria, such as the influenza bacillus and the meningococcus, have repeatedly been found in encephalitic foci. The seat of acute encephalitis may be in the cerebral substance, in the basal ganglia, in the ventricular region or in the medulla oblongata, and the clinical pictures produced vary in accordance with the distribution.

Symptomatology.—Acute encephalitis of the cerebrum may manifest itself in a variety of ways. In a certain proportion of the cases it is characterized by severe brain symptoms and high fever. The child is extremely ill and may lie motionless for hours or days as if life were already extinct; there may be tetanic spasms and general convulsions, rigidity of the neck, opisthotonos, hyperpyrexia up to 41° C. (105.8° F.). The relatives usually state that the disease has been diagnosed meningitis and that a fatal prognosis has been given. Nevertheless, an attentive observer will note even in a case of this kind, certain symptoms pointing to a localized disease of the brain. There may be tremor of one or both extremities on one side; facial palsy, monoplegia in one extremity, aphasia or sensory disturbances frequently develop suddenly. The eyes are often in a position of permanent convergence, the eyeballs directed upward; after a certain time conjugate deviation ensues ("the patient looks at the disease focus"). Optic neuritis is not infrequently present. The child continues in this condition for a few days or a week. If, to the great astonishment of the relatives, the child survives, paralytic symptoms usually remain which gradually give the disease the character of a cerebral infantile palsy. Unilateral or general convulsions or idiocy may also remain as the result of the encephalitis.

The **course** of infantile encephalitis is not always as violent as has just been described. Sometimes the fever is moderate; vomiting, headache, and rigidity of the neck are overshadowed by the irritative and paralytic phenomena in the extremities and in the cranial nerves. The inconstancy of the symptoms and frequency of localized and general tremors are quite characteristic of these subacute forms of encephalitis.

Finally there are cases with an insidious course, presenting a symptom-complex which resembles that of brain tumor. Oppenheim has called attention to cases of this kind, which are usually regarded as cases of brain tumor until the favorable outcome casts a doubt on the diagnosis. Of course, the diagnosis of encephalitis in a case of this kind must be tentative; but we practically know of no other slowly progressive morbid processes in the brain that end in such complete recovery as inflammatory, or at least vascular disturbances. Recovery may be complete, or local and general cerebral symptoms may persist. In a case under my own observation in which there had been bilateral paralysis of the abducens and choked disc, permanent epilepsy resulted, and it is conceivable that a circumscribed inflammation in a silent region of the brain might end in recovery and leave the patient an epileptic. It is possible that cases of acute neuritis ending in recovery with neuritic atrophy also belong to this category.

When the inflammation is situated in the mesencephalon or medulla oblongata instead of in the hemispheres, it gives rise to certain clinical conditions which deserve special description.

Wernicke first described a symptom-complex which has since been designated *acute hæmorrhagic superior poliencephalitis.* A short prodromal period, marked by headache, vomiting and vertigo, is followed by hebetude, rigidity of the neck, ocular palsies, disturbance of the gait and ataxia. As a rule there is no fever. The course of the disease is progressive. Death usually occurs in from 1 to 2 weeks. The causes of this condition are believed to be alcoholism and infectious diseases (influenza). Post mortem, acute hæmorrhagic inflammation of the gray matter in the third ventricle and in the aqueduct of Sylvius, extending over into the fourth ventricle, has been observed. In children this form of encephalitis is rare.

When the acute inflammatory process is situated in the central portions of the medulla oblongata nearer the spinal extremity, the symptoms are those of an *acute inferior poliencephalitis.* The bulbar nerves, that is, the facial, hypoglossus, vagus, and spinal accessory, are chiefly involved. Disturbances of speech, dysphagia, interference with the movements of the mouth and tongue, aphonia, disturbance of the pulse and respiration, dribbling of saliva, hysterical laughing, and crying are present. It is needless to say that if the inflammation is so extensive as to involve all the nuclei of the cranial nerves, the symptoms of a superior and inferior nuclear palsy are combined; and if the process also involves the pyramidal tract, hemiplegia, monoplegia, tremor and intention tremor develop. A severe inflammation of this kind affecting the bulbar region, which occurs chiefly after infectious diseases and as the result of poisons but may, like poliomyelitis, develop spontaneously, usually terminates fatally, recovery with permanent symptoms being a rare event (Kollarits). It is worthy of note, however, as showing the analogy with doubtful forms of encephalitis resembling brain tumors, that even an acute bulbar affection may run a mild course and end in complete recovery (as for example, after meat and sausage poisoning). Whether in cases of this kind inflammation or changes resembling inflammation are actually present is difficult to decide, particularly as acute fatal bulbar affections sometimes exhibit no post-mortem lesion except possibly aggregations of micrococci in the portions of the brain supposed to be diseased.

Finally, we should devote a special paragraph to *disseminated myeloencephalitis.* In this affection, multiple inflammatory foci are produced in the brain, medulla oblongata and spinal cord, which may run an acute, subacute or chronic course. The cause is in all probability to be sought in the infectious diseases. The symptoms are extremely variable consisting as they do in a combination of symptoms referable to the cranial and bulbar nerves and disturbances in the extremities grouped without any apparent order. Either the cerebral or the spinal symptoms may predominate, and the clinical picture may be chiefly that of a cerebral infantile palsy, of a bulbar affection or of a poliomyelitis. A number of observers have contributed post-mortem proofs of this widespread disease, which extends over the entire central nervous system (Redlich, Schupfer and others). The terminal stage of the disease is a condition in which disturbance of speech and of the intelligence, tremor and spasm are the most prominent symptoms. The picture of a multiple sclerosis may thus be simulated. Indeed, it is not too much to say that most of the cases diagnosed as multiple sclerosis in childhood represent the remains of circumscribed inflammations in the brain and spinal cord which have ended in recovery.

The **differential diagnosis** from multiple sclerosis is based on the history of some infectious diseases, the acute onset, and on the arrest of symptoms or, in other words, the failure of the disease to progress steadily. Post mortem the remains of a former inflammation are found instead of, as in multiple sclerosis, the formation of neurogliar tissue exclusively.

In view of the great variety in the different forms of encephalitis it is manifestly impossible to lay down hard and fast principles for making a diagnosis. The difficulties appear still greater when we remember that hereditary syphilis is also capable of producing in the brain substance alterations which clinically resemble the various forms of encephalitis very closely. Hence, in order to avoid unnecessary repetition, it seems wiser to omit any discussion of the differential diagnosis of encephalitis at this place and to refer the reader to the various diseases which must be considered in that connection, namely, meningitis, hereditary syphilis, brain abscess, sinus thrombosis, embolism, the various forms of bulbar palsy, cerebral infantile paralysis, brain tumor and multiple sclerosis. It should be emphasized, however, that in any disease beginning acutely with fever and presenting, in addition to general symptoms of brain irritation, rapidly developing and persistent focal symptoms, the possibility of acute encephalitis should be borne in mind. The diagnosis is confirmed if the symptoms of cerebral palsy persist after the acute stage. At the beginning of the disease lumbar puncture is useful for the purpose of excluding meningitis. But in spite of the greatest care in diagnosis there will always be found cases that run such a rapid course, with hyperpyretic convulsions, that death ensues before the cerebral localization of the disease can be definitely established.

The **prognosis** of acute encephalitis is always extremely grave. Even if the child survives, some form of permanent palsy may be expected. However, if it is a question between meningitis and encephalitis, a decision in favor of the latter diagnosis is more favorable for the patient since it offers some ray of hope, whereas meningitis of equal severity would be hopeless. The possibility of complete or practically complete recovery has been discussed. The usual termination of an encephalitis that ends in recovery is a cerebral infantile palsy.

The **treatment** of an acute encephalitis has for its object to deplete the blood vessels of the brain, control the fever, support the heart, and, finally, combat the brain symptoms. The first requisite is absolute rest in a darkened room, to protect the patient against light and loud noises. An ice bag is applied to the head or, better, a Leiter's coil or rubber cap through which a constant stream of cold water is passed, even at night. Direct depletion with leeches is strongly to be recommended, selecting for the application of the leeches the mastoid process on the side which is suspected to be the seat of the disease. Venesection, which is employed in adults in cases of severe congestion of the brain, should be considered in children only in exceptional cases. Depletion through the intestinal tract should always be tried, calomel being the best remedy for that purpose. With a similar object in view warm foot-baths may be given in the hope of altering the distribution of the blood.

Cold packs and baths, repeated at short intervals, are the principle means employed to combat the fever; although quinine, antipyrin or aspirin may have to be administered either by mouth or by rectum in order to reinforce the antipyretic effect of these measures. Hot baths which are commonly employed in the treatment of cerebrospinal meningitis have occasionally been employed with good results in acute encephalitis.

The most important measures for supporting the heart are those which diminish the fever and combat the violent brain symptoms. Another important factor in this respect is the administration of sufficient nourishment even if this be no more than milk, eggs and soup. If the child refuses to take nourishment it must be administered with a spoon like medicine, employing a mixture of sugar, egg, milk, milk of almond or a little somatose, which can usually be done without any difficulty. Alcohol is not to be recommended; coffee, tea and cardiac stimulants are more advisable.

Severe brain symptoms such as convultions, jactitation, insomnia may be combated with large doses of bromides and chloral hydrate. Lumbar puncture, unless for diagnostic reasons, is not to be recommended in a pronounced case of encephalitis.

After the acute stage has subsided, potassium iodide is usually administered for some time.

For the treatment of any resulting paralysis or epilepsy, see under cerebral infantile palsy

II. BRAIN ABSCESS

Brain abscess is a not uncommon disease in childhood.

The causes are head injuries, diseases of the ears, nose or other structures of the head, suppuration in distant organs and general sepsis.

We accordingly distinguish traumatic, otogenic, rhinogenic and metastatic abscesses. When no cause can be discovered, the term idiopathic abscess is used. Abscesses occurring as sequels of suppuration or acute inflammatory diseases of the meninges or of the blood vessels must be regarded as symptoms of this disease and therefore require no further discussion in this place.

With regard to the frequency of brain abscess in childhood, Gowers found that of 223 cases, 24 occurred during the first, and 48 during the second decade of life. Holt collected 27 cases, most of them in infants, although during the earliest years of life the tendency is rather toward general suppurative meningitis than toward brain abscess. Of the various forms of brain abscess the otogenic and traumatic are the most common in childhood. "Idiopathic" abscesses not infrequently occur in children and are probably due to a former unrecognized septic process.

Pathology.—Brain abscess is the result of acute inflammation of the brain. The tissue in the centre of a diseased focus breaks down or undergoes rapid suppuration: the resulting cavity is at first irregular in outline and contains greenish yellow, fetid pus, masses of necrotic brain tissue, sometimes fluid, and always bacteria. In addition to the pus producing organisms—streptococcus and staphylococcus—there are found, among others, pneumococcus, pyocyaneus and in a few cases the tubercle bacillus (Fränkel). The abscess grows very rapidly and may be as large as a pea or occupy an entire hemisphere. After a time, (from two to three weeks in traumatic abscesses, Lebers) the pus cavity becomes surrounded by a membranous capsule, which is usually smooth and vascular and varies in thickness up to 5 millimetres according to the duration of the abscess. As the abscess becomes encapsulated it assumes a more spherical shape. The development of a capsule does not necessarily imply that the disease has become arrested, for it may remain latent for a long time (28 years in the case of traumatic abscess, Nauwerk). Otitic abscesses remain latent at most 1½ years (Macewen). Not infrequently the abscess connects by a fistula with the surface of the brain or a diseased portion of the skull. The brain substance in the immediate neighborhood of the abscess usually exhibits inflammation and softening; large abscesses cause pressure symptoms in distant portions of the brain, flattening of the convolutions and internal hydrocephalus.

The different varieties of brain abscess present certain special features. *Traumatic* brain abscess is almost always solitary and is usually situated in the cerebrum, more rarely in the cerebellum. It may remain latent for years. *Otitic* brain abscess occurs chiefly as the result of chronic suppurative catarrh of the middle ear (cholesteatoma, polypi), the morbid process beginning in the bone and involving the dura secondarily. It is situated in the temporal lobe (usually on the right side, Körner), or in the cerebellum. According to Körner 82 per cent. of otitic brain abscesses in children occur in the cerebrum and 10 per cent. in the cerebellum. In adults the proportion is 63 per cent. in the cerebrum and 37 per cent. in the cerebellum. The abscess is usually solitary and may attain a considerable size, particularly in the temporal lobe. The abscesses which rarely follow suppurations in the nose and in the orbital cavities are usually situated in the frontal lobes. *Metastatic* abscesses are particularly frequent in children. The primary disease may be a putrid affection of the lungs (bronchiectasis, gangrene of the lung, sometimes ulcerous tuberculosis), more rarely suppurative peritonitis; thrush even is given as the cause in few cases (Zenker. Wagner). Metastatic abscesses are almost always multiple. They exhibit a predilection for certain portions of the brain and, in addition to putrid pus, occasionally contain some of the structural elements of the primary focus (pigment from the lungs). So-called *infectious* abscesses are sometimes seen after cerebrospinal meningitis, typhoid fever and influenza and have a similar significance, In a good many of these cases an otitis probably represents the connecting link. The occurrence of primary *idiopathic* brain abscess is quite properly doubted by many authorities (Huguenin, v. Bergmann, Broca and others), particularly as a long interval of time may elapse between the primary suppuration and the appearance of the abscess. Martius believes that a primary abscess may be produced by a bacterial cause of cerebrospinal meningitis.

The clinical phenomena vary according to the case, the seat of the abscess and the stage of the disease. We distinguish: (1) the initial stage; (2) the stage in which the abscess is well developed, and (3) the terminal stage. In addition there is in many cases a latent stage, which occurs between the first and second and must be inferred chiefly from the history (Macewen, Oppenheim).

The initial stage is characterized by headache, vomiting, chills and general prostration, with fever of variable degree. These symptoms may easily be overlooked in the presence of a head injury or a suppurative otitis, which in themselves produce the picture of severe illness. The initial stage may last from 1 to 6 days. It is followed by a period of latency during which either all symptoms are entirely absent ("pure latency") or the patient may be in comparatively good health, occasionally interrupted by headache, fever, a tendency to drowsiness or sudden chills ("impure latency," Oppenheim, Huguenin). Optic neuritis may also be present. Leaving out a few exceptional cases, this period rarely lasts longer than a few months. The active or manifest stage of the disease either follows directly upon the initial stage or may be separated from it by the latent period. It is during this stage that the physician often sees his patient for the first time. The general symptoms of the first stage are still present in more or less characteristic form and, in addition, other signs referable to the seat of the disease make their appearance. The headache, which is usually less intense than during the initial stage, is often referred to a definite part of the head and thus affords some clue to the seat of the brain abscess, although great caution is necessary in this respect in the case of children. It is increased by anything which tends to raise the blood pressure (contraction of the abdominal muscles or coughing). Sometimes percussion of the skull elicits distinctly localized pain. Vomiting is more marked in cerebellar abscess and often follows a change to the erect posture. Convulsions are more frequent in children than in adults. The pulse rate is distinctly reduced and may fall as low as 30 to 60 beats in the minute (Macewen, Baginsky, Gluck). The pulse is usually irregular and intermittent. Cases are observed, however, especially in children, in which the entire active stage of the disease is marked by increased pulse frequency. Respiration may be slowed in cerebellar abscess, and Cheyne-Stokes breathing is occasionally observed. The temperature is usually normal and often subnormal, an important point in the differential diagnosis from suppurative meningitis and sinus phlebitis. Psychic changes which often occur are interesting. They consist in loss of memory, inability to concentrate, sluggish reaction to stimuli. In smaller children there may be stupor and delirium, with violent headache as the dominant symptom. In addition to these general symptoms many, but by no means all cases exhibit focal symptoms which afford a clue to the seat of the abscess. It is characteristic of brain abscess that the abscess may attain a considerable size before it produces any localized symptoms. The reason of this lies in the slow growth of the abscess, the gradual encroachment on the brain substance, and the fact that the more delicate portions of the brain, the internal capsule and the medulla oblongata, are rarely the seat of abscess. Small multiple abscesses particularly of the metastatic variety rarely cause paralytic symptoms.

The focal symptoms may be due directly to the seat of the abscess or to pressure on distant portions of the brain. Abscesses in the temporal lobe, which must be thought of after disease of the middle ear, are characterized by word deafness (the words are heard but their meaning is not understood) and loss of hearing in the opposite ear. In addition to these symptoms, which are difficult to elicit in children, we have as the result of pressure on the internal capsule paralysis of the facial nerve or the extremities on the opposite side, and from pressure on the oculomotor, which runs along the base of the brain, paralysis of that nerve on the same side (rarely of the internal muscles of the eye). When the abscess is on the left side, a purely motor aphasia from pressure on the third frontal convolution is present. Abscesses situated in the frontal lobe rarely produce definite focal symptoms; although, under the same conditions as obtained in abscess of the temporal lobe, hemiplegia of the other side of the body and aphasia may result as distal effects. An abscess in the parietal lobe, particularly one of traumatic origin, may lead to cortical convulsions and hemiplegia on the opposite side of the body, which often attacks the individual extremities in succession, with convulsions and fever. Abscesses in the occipital lobe are rare; they lead to hemianopsia, but the symptom is not reliable. In abscesses involving both the occipital lobes blindness is observed. The characteristic symptoms of cerebellar abscess are rigidity of the neck, interference with the gait, vomiting, violent headache, and distal symptoms referable to the corpora quadrigemina, the pons and the medulla oblongata, with paralysis of the ocular muscles, the bulbar nerves, hemiplegia of the opposite side or paraplegia. These disturbances are observed chiefly when the lesion involves the worm; when the abscess is situated in the cerebellar hemispheres, local disturbances may be absent for a long time. In the pons and medulla oblongata, abscesses are very rare (Cassierer).

This stage of manifest symptoms, which is characterized by a great variety of morbid phenomena, may merge directly into the *terminal* stage by a steady increase in the symptoms; or the terminal stage may follow immediately upon the period of latency. The pus may find its way to the surface of the brain and set up acute purulent meningitis, with convulsions, chills, acceleration of the pulse, fever and disturbances of the respiration, or the abscess may rupture into the ventricles of the brain (pyocephalus), in which case the course is exceedingly violent and the above-mentioned symptoms are very intense. The patient goes into collapse almost at once and death ensues a few hours later. In rare cases the terminal symptoms are less pronounced, so that a diagnosis of "atypical" meningitis is made. Again, the brain symptoms may be entirely absent and death may occur from general marasmus and pyæmia.

In order to establish the **diagnosis** of brain abscess there must be, as Oppenheim quite properly contends, an exciting cause. In the case of children, however, it is often difficult to find the cause as diseases of the ear and nose frequently remain latent, and a long interval often intervenes between an injury to the head and the appearance of brain abscess. Conversely, pus-retention following a head injury or an acute otitis may produce brain symptoms which promptly subside after the pus has been evacuated by surgical intervention. Even if the diagnosis of intracranial suppuration is positive, there still remain to be considered purulent meningitis and sinus phlebitis. Purulent meningitis is quicker to develop after an injury or the onset of an acute otitis then is the case with brain abscess, which requires several days or weeks to form. Acute brain symptoms such as convulsions, delirium and coma are more intense from the beginning and are constant; the local symptoms are more evanescent. The fluid obtained by lumbar puncture, which may be performed even if abscess is suspected, contains pus corpuscles. The diagnosis of serous meningitis, which may also occur after otitic affections, is more difficult. Its symptoms are such, however, that there is more danger of confusing it with purulent meningitis than with brain abscess. Sinus phlebitis is characterized by a typical pus-temperature, with chills and remissions; the only positive symptoms are those referable to the blood vessels, which will be discussed in another place.

If the case is one of chronic brain abscess, the possibility of a brain tumor must be considered, particularly as in scrofulous and tuberculous individuals the association of brain tubercle with discharge from the ears is quite within the bounds of possibility. The absence of the initial rise of temperature, the more localized character of the focal symptoms and distinctly slower course of the disease, the presence of marked choked disc are in favor of brain tumor. The probable seat of the suspected neoplasm is of some importance, as abscess is rarely situated in the pons or in the medulla oblongata, while, on the other hand, brain tumors in children are not infrequent in these regions. If, in the presence of an aural affection, symptoms referable to the temporal lobes are detected, this is in favor of brain abscess. If the brain symptoms persist for some time without undergoing any change and the general condition is not favorable, the probabilities are rather in favor of an abscess. The sudden occurrence of the terminal stage, when symptoms of brain tumor have been present only a short time, is quite characteristic of abscess.

Even with the aid of all these points in the differential diagnosis we cannot always avoid mistakes, or at least it may have to be admitted in individual cases that the diagnosis is impossible. The fact that abscess shows a tendency to be multiple and a preference for the less characteristic portions of the brain renders its recognition more difficult. In a case of doubt between tumor and abscess it is better, in the interests of the patient, to adopt the latter theory and, if possible, to attempt operative interference.

Although brain abscess may remain latent for a long time, and although it may rarely undergo absorption or rupture on the exterior of the brain, the **prognosis** is distinctly unfavorable. In any case of brain abscess the possibility of rupture, either spontaneous or from traumatism, followed by sudden death must be borne in mind.

For all that in many cases the diagnosis of a brain abscess may mean the patient's salvation because it offers the chance of a successful operation, whereas in cases of meningitis or brain tumor the chances for successful operation are much fewer. We must not omit to emphasize the importance of prophylaxis in brain abscess, which consists in the careful treatment of all suppurative wounds of the head and aural affections. The results of the many operations on brain abscess which have been performed in recent times are not altogether unfavorable. (Oppenheim computes 36 recoveries out of 53 cases of traumatic abscess, Körner 51 out of 92 cases of otitic abscesses.) Even infants and very small children may recover (Holt had 5 successful cases with operation). Without going into the technic of the operation it may be stated that the tendency nowadays is toward free opening of the skull and, if the abscess does not at once come into view, fearless exploration of the suspected portions of the brain with a needle. The favorable prospects of operation are unfortunately marred by the fact that abscesses are prone to be multiple so that, after a large abscess has been evacuated, death may ensue from a second purulent focus, the existence of which had not been suspected. Evacuation of the abscess without thorough treatment directed against the primary trouble is of course of no more value than any other symptomatic treatment. If no operation is performed, the patient is very likely to die sooner or later from the effects of the abscess; hence, if the diagnosis is half way positive, operation should be attempted in spite of the uncertainty of its results.

III. SINUS THROMBOSIS

Thrombosis of the cerebral sinuses may occur as the result of grave debilitating diseases ("marantic" or "primary" thrombosis) or may accompany some acute inflammatory affection of the cranial structures ("phlebitic" or "secondary" thrombosis).

Anatomy.—The superior longitudinal sinus follows the sagittal suture along the top of the skull to the torcular Herophilii, situated at the occipital prominence. At this point it divides into several other sinuses; the occipital sinus, which passes downward and empties into the occipital vein; the straight sinus, which passes forward and, after receiving the blood from the vena magna Galeni which drains the chorioid plexus, empties into the inferior longitudinal sinus (falciformis minor) which courses along the lower border of the falx cerebri; the two transverse sinuses on each side, which pass close to the petrous portion of the mastoid bone and empty directly into the jugular veins. The transverse sinus in front gives off the inferior petrous sinus between the clivus and the petrous portion of the temporal bone, and the superior petrous sinus at the upper border of the petrous portion; these, after passing around the sella turcica unite to form a large channel, the cavernous sinus, which communicates with the ocular veins. Besides the venous trunks that have been mentioned, the sinuses, especially the sinus of the falx cerebri, communicate with numerous small veins that perforate the skull at various points. The cerebral sinuses represent cavities of varying width and outline, provided with small valves; they contain minute septa and villi which encroach on the lumen and explain their special tendency to thrombosis.

Marantic sinus thrombosis results from conditions characterized by marked loss of fluid or weakness of the heart, such as severe intestinal catarrah, long-continued suppuration, endocarditis, myocarditis, syphilis and tuberculosis with severe cachexia.

The most frequent seat of the thrombosis is the longitudinal sinus and next to that the transverse sinus. The process may involve most of the cerebral sinuses. The thrombi are at first of a reddish color, later more yellowish, of a loose consistency and very friable; the attachment to the wall of the sinus, which is but little altered, is at first loose but gradually the clot grows firmly adherent.

Fig. 43.

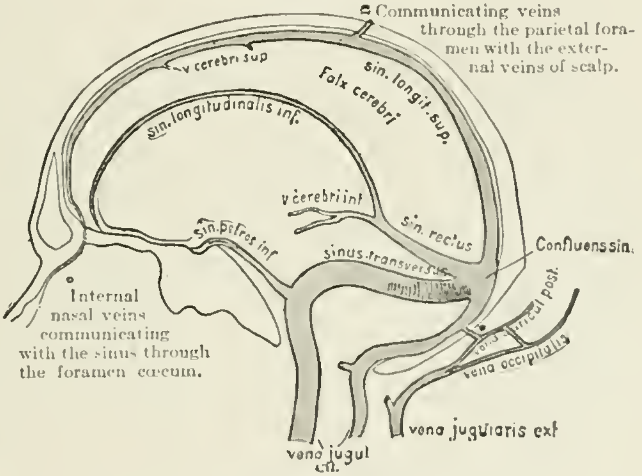


Diagram showiog the relations of the superior longitudinal and transverse sinuses to external veins. (*) After Leube.

Secondary (phlebitic) sinus thrombosis results from disease of the ear, disease of the bones, suppuration within the cranial cavity, the orbits and suppurative processes in the face (furuncle, erysipelas).

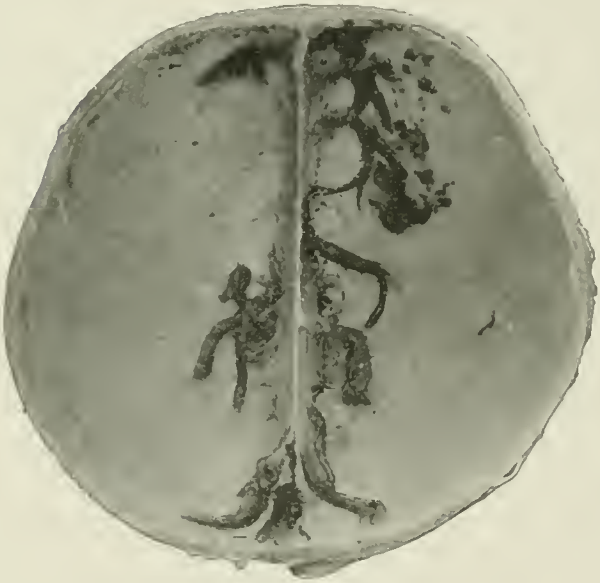
Sinus thrombosis due to diseases of the ear may be situated in the transverse sinuses, especially on the right side, the petrosal or cavernous sinuses. A phlebitic thrombus is discolored, diffluent and in a state of decomposition, and the sinus wall exhibits the signs of acute inflammation, which may extend to the neighboring veins. The occlusion of the sinus and the spread of the inflammation may produce marked changes in the meninges and in the brain. Hyperæmia, local or diffuse meningitis, extensive meningeal hæmorrhages, areas of softening, hæmorrhages, brain abscess and hydrocephalus are observed.

The bacteriologic findings are positive not only in marked phlebitic but also in marantic thrombosis (Heubner). When it is remembered that in the latter form of sinus thrombosis also the primary disease is usually due to bacterial toxins, Marfan's refusal to accept the theory of a purely "marantic" thrombosis appears quite justifiable: according to him, every sinus thrombosis is to be regarded as septicæmic in character, and he distinguishes only between those forms which are due to a local cerebral lesion and those which result from general diseases. From this point of view we are justified in classifying sinus thrombosis among the inflammatory diseases of the central nervous system.

The **symptoms** of sinus thrombosis may be completely masked by the symptoms of the existing disease. This is particularly the case in so-called marantic thrombosis, because the convulsions or loss of consciousness to which they give rise are very apt to be regarded as the forerunners of death.

The *cerebral* symptoms are not very characteristic; they consist in headache, vomiting, crying out at night, convulsions, somnolence and coma. In addition, there may be strabismus, nystagmus, dilatation of the pupils and, more rarely, palsies in other parts of the body. In an infant the sudden occurrence of convulsions followed by somnolence is very suggestive of sinus thrombosis.

Fig. 44.



Sinus thrombosis in a child of two years and four months.

The temperature is of considerable diagnostic importance. In septic sinus thrombosis it may attain extreme degrees and also be characterized by repeated chills. If, in addition to a sudden extreme rise of temperature, the above-mentioned cerebral symptoms are present, and if this occurs in the course of a purulent otitis, for example, the change in the patient's condition may be sufficiently definite to suggest the onset of sinus disease.

Local œdema and the presence of secondary thrombi from extension are important signs for the diagnosis of a sinus thrombosis and afford direct information in regard to the character and seat of the disease. Thus, thrombosis of the transverse sinus produces œdema behind the mastoid process and, according to Jansen, not infrequently also thrombosis of the upper portion of the jugular vein; the thrombosis in the latter can sometimes be felt or may betray itself by pain when the head is moved, by persistent lateral inclination of the head and by dysphagia.

By making pressure in the jugular foramen it may cause disease of the nerves which pass through the foramen—the vagus, the spinal accessory and glossopharyngeal—and bulbar symptoms, and may indeed be the direct cause of death. Occlusion of the longitudinal sinus is followed by swelling of the veins in the skull and in the scalp. Thrombosis of the cavernous sinus may conceivably produce swelling of the eyelids (which may be unilateral), inflammation of the orbital contents, disturbance of the occular muscles and trifacial neuralgia. These local symptoms are, however, frequently absent and cannot therefore be relied upon for the diagnosis. In many cases a general pyæmia may be the only sign of an existing phlebitis and the cerebral symptoms may be quite inconspicuous.

The **course** of sinus phlebitis is almost always rapidly fatal. Rarely, particularly in the marantic form, the disease may last several weeks before death occurs. Complications due to secondary disease, particularly pulmonary embolism, may occur. Recovery is extremely rare and, owing to the uncertainty of the diagnosis, its frequency is difficult to estimate. There is, however, a possibility of a collateral circulation being established (Hölscher), or the sinus may become obliterated by fibrous tissue with secondary hydrocephalus (Marfan), constituting a temporary recovery, as has been proven by subsequent autopsies. According to Fischer inflammatory disease of the sinuses ending in recovery is one of the causes of cerebral infantile palsy.

Operation should always be considered, particularly in cases of phlebitic sinus thrombosis. In cases of otitis media the operator often finds, on opening the mastoid process, that the lateral sinus is thrombosed and thus performs the operation, although he had not originally intended to do so. Some surgeons combine the operation for sinus thrombosis with ligation of the jugular vein. Numerous results have already reported from this operation and they are not bad; 58.4 per cent. of recoveries according to Körner. For the technic of the various methods of operation the reader is referred to Körner and Jansen.

IV. THROMBOSIS AND EMBOLISM OF THE CEREBRAL VESSELS

Occlusion of the arteries in the brain is far more rare in the child than in the adult because of the absence of arteriosclerosis, which is an important etiologic factor in later life. In hereditary syphilis arterial occlusion may result directly from the vascular disease. In childhood primary thrombosis of the brain vessels is rare as compared with sinus thrombosis, as the latter develops in the form of marantic sinus thrombosis under conditions such as exhausting disease, lowering of the blood pressure and the like, which in the adult usually produce thrombosis of the cerebral arteries. Sometimes occlusion of the cerebral vessels occurs secondarily from extension of a sinus thrombosis. Embolism of the cerebral vessels is more frequent and is always due to some inflammatory disease within the vascular apparatus. The acute infectious diseases, such as diphtheria, scarlet fever, pneumonia and even measles (Baginsky), may be followed by cerebral embolism, the cerebral occurring as a late complication of the primary trouble. After rheumatic endocarditis occlusion of the cerebral arteries is rare. Poisoning and burns are also mentioned among the causes of cerebral embolism in the child.

The **pathologic findings** vary with the interval that has elapsed between embolism and death. In recent cases hæmorrhages or acute hæmorrhagic inflammation, so-called "red" softening, is found. This is followed by "yellow" softening, due to changes in the blood-pigment and the breaking down of tissue. If the site of the embolism is not well supplied with blood, the area of softening is white in color, so-called "white" softening. The final result of this encephalomalacia is absorption, scar formation, cyst or sclerosis. Even in the later stages the relation to the blood vessels is easily recognized.

It follows from the above description that the dividing line between infectious embolic encephalomalacia and primary encephalitis cannot always be sharply drawn pathologically, particularly in the later stages of the disease. As a matter of fact, the only difference between the two conditions lies in the size and number of the embolic plugs that are carried to the brain.

The characteristic feature of cerebral embolism is the sudden occurrence of grave brain symptoms. The disease is usually ushered in by convulsions; this is followed by a stage of coma, during which local paralytic and irritative symptoms are often recognized. Sometimes the disease is preceded by general cerebral symptoms, headache, restlessness, vomiting and hebetude. An accurate diagnosis cannot be made until focal symptoms make their appearance, the most important of which are hemiplegia, aphasia and sensory paralysis. As the initial symptoms subside, the localizing signs become more distinct and afford some clue to the identity of the occluded artery (see diagram under brain tumor). A knowledge of the parts of the brain which are supplied by the various cerebral arteries, as shown in the following table, will aid in localizing the lesion:

Arteria cerebri anterior s. corpor. callosi: Pons, median surface of frontal and parietal lobes.
Art. cerebri med. s. Fossæ Sylvii: Basal ganglia, part of the internal capsule, cerebral cortex.
Art. chorioidea: Chorioid plexus.
Art. cerebelli inferior: Under surface of cerebellum.
Art. cerebelli anterior: Cerebellum.
Art. cerebelli superior: Upper surface of cerebellum.
Art. cerebri posterior: Occipital lobe, posterior portion of basal ganglia.

Embolism of the artery of the fissure of Sylvius, which is followed by the appearance of hemiplegic symptoms, is the most common form. If a small end artery is occluded, marked symptoms of paralysis may be wanting.

When arterial embolism involves a large area of the brain or affects a vital portion, it may prove rapidly fatal. The child does not regain consciousness after the initial coma; profound collapse and increasing heart weakness develop. More frequently the course of the disease is favorable; the initial symptoms gradually subside and a palsy results which presents the picture of cerebral infantile paralysis. In such a case the occurrence of brain embolism can only be inferred from the history.

Diagnosis.—The presence of a cause and the history of sudden onset are essential. But since in childhood various cerebral affections begin with convulsions, the distinction from a circumscribed meningitis, a cerebral hæmorrhage or an encephalitis may at first present great difficulties and in some cases may be impossible.

Treatment of the acute attack consists in the main in supporting the heart, and mitigating the severity of the cerebral symptoms (see treatment of encephalitis). In old cases the treatment is the same as that of cerebral infantile paralysis.

V. CEREBRAL SCLEROSIS

The term cerebral sclerosis is used to describe a number of pathologic conditions characterized macroscopically by thickening, contraction and brownish discoloration of the brain substance, microscopically by proliferation of the connective tissue, particularly in the septa of the brain, and in the perivascular tissue, and by thickening of the vessel walls. The degree to which the nerve tissue is involved in the process varies; ganglion cells and nerve fibres may atrophy or degenerate and disappear completely. Sclerosis may affect only certain portions of the brain, forming hard knotty tumors (tuberous sclerosis), or an entire lobe (lobar sclerosis), or it may cause shrinking and atrophy of an entire hemisphere either of the cerebrum or of the cerebellum (hemispherical form of sclerosis).

These forms of sclerosis merely represent the terminal conditions of some severe cerebral process and may accordingly be due to a variety of causes. They occur as the result of intra-uterine diseases (combined with porencephaly); they may be the products of brain softening due to hereditary syphilis, or represent the remains of an inflammatory disease of the brain, etc. Accordingly, the clinical symptoms which mark the onset or subsequent course of these sclerotic conditions are by no means uniform. In the beginning they frequently resemble the symptoms of a congenital paralysis, encephalitis, brain embolism, etc.;

later in the course of the disease the variegated picture of cerebral infantile palsy is simulated.

Hence but little is gained by collecting these various forms of sclerosis into a common group, be it never so comprehensible (Richardiere) nor is the subject of cerebral disease in children thereby rendered more intelligible. A better plan is to analyze the causes of sclerosis and subdivide them into various groups, according to the etiology and clinical symptoms. It should be noted, however, that the central nervous system in childhood exhibits a peculiar tendency to react to morbid processes by the formation of fibrous tissue.

Conspicuous in this group of anatomical findings is a condition which has been greatly studied in recent years and has received the name of *diffuse cerebral sclerosis* (Strümpell, Heubner, Busse, Schmaus, Bullard, Hugo, Weif, Frankl-Hochwart and others). The symptoms of the disease are not present at birth, but develop later "often in the midst of perfect health," although the child may have previously exhibited some defect in its mentally or bodily development. Diffuse sclerosis is especially apt to attack children in the first years of life, older children and adults are rarely affected.

The **causes** of diffuse cerebral sclerosis are not known. Syphilis, parental alcoholism, and traumatism are mentioned. In one case of Pfaundler's (private communication) syphilis, parental consanguinity, and alcoholism and beginning paralysis in the father were present.

The **symptoms** of the disease are both psychic and somatic.

As a rule the first thing that is noticed is a diminution of the intelligence, a sluggishness in the child's movements, a loss of interest in its surroundings. This psychic change may be considerably increased in the subsequent course of the disease and complete idiocy may ultimately develop. The power of speech is rapidly lost and the child expresses itself only in grunting, inarticulate sounds. There are cases in which the intelligence remains approximately normal and is indeed retained until death. Among the bodily symptoms awkwardness and sluggishness in the movements of the body are first noted. The muscular tone is increased; the muscles are tense and boardlike; rigidity soon develops and ultimately leads to contractures. The muscular rigidity is not confined to the extremities but attacks the muscles of the face and the muscles of mastication as well. Sometimes the legs are first involved, in other cases the rigidity appears in all the extremities at the same time. At the height of the disease the child, owing to the spastic contractures, is deprived of all power of motion; the arms are flexed and pressed against the trunk, the legs are extended and crossed; there are distressing convulsions affecting the muscles of the jaw and general muscular twitchings which may be rhythmical, and absolute inability to carry out any intentional movements. Tremor, ataxia and athetosis are not infrequently present. True paralysis and atrophy do not occur; but the children rapidly emaciate during the subsequent course of the disease.

Among ocular symptoms, nystagmus, strabismus, conjugate deviation, choked disc and atrophy of the optic nerve are observed. The pupils react promptly to light and accommodation.

Disturbances of deglutition, phonation and the movements of the tongue are present in rare cases.

The course of the disease is occasionally interrupted by apoplectiform, rarely epileptic attacks, which leave the child in a much worse condition. Death ultimately results from decubitus, pneumonia or some other terminal condition.

The above-described symptoms are by no means present in every case. Many of the symptoms may be wanting and, on the other hand, the course is not always steadily progressive as here described. It must be remembered that the clinical picture is as yet but imperfectly defined and an accurate description of the symptoms cannot be thought of.

Such pathologic observations as have been made in diffuse sclerosis are, in the main, quite uniform. Not only the brain, but the medulla oblongata and the spinal cord as well, is diffusely thickened and hardened, so that cross-sections retain their sharp edges which is not usually the case in the brains of children. The hardening and thickening process is particularly marked in the white substance and in the basal ganglia. The color of the medullary substance is yellowish white, that of the cortex a pale gray. There are no areas of special thickening. Histologically a considerable increase of the connective tissue and of the medullary cells is found, while the ganglionic elements are intact. According to Schmaus, the pathologic picture represents the end-product of an interstitial inflammatory process and his view has been accepted by most authors.

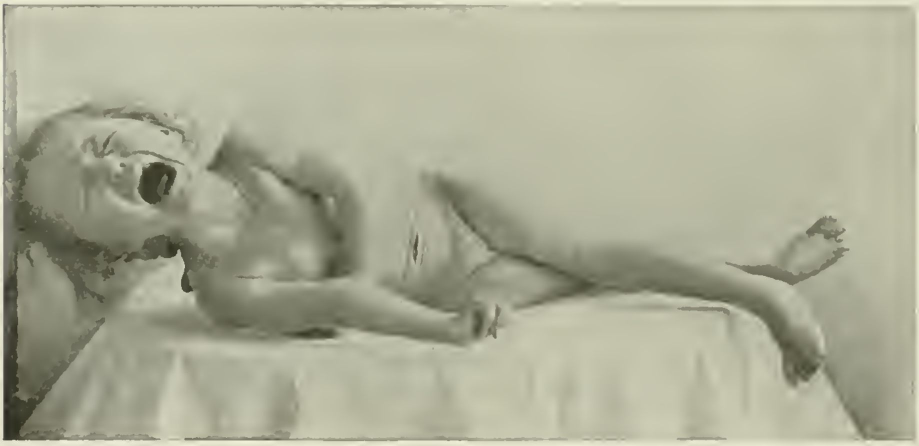
The term pseudosclerosis is applied to a condition characterized by symptoms such as are usually ascribed to multiple sclerosis—disturbance of the speech, tremor, nodding of the head and disturbance of deglutition—and by complete absence of all pathologic findings. In children pseudosclerosis appears to be an endogenous disease (Frankl-Hochwart).

VI. POLIOMYELITIS

Poliomyelitis is an acute inflammation of the spinal cord which chiefly attacks the distribution of the anterior central artery (Kadyi) in the gray matter of the anterior horn. The term poliomyelitis which was introduced by Kussmaul is therefore quite appropriate (*πόλιος*. gray), but it does not imply a sharp distinction from myelitis (see below).

The investigators of poliomyelitis had to overcome two great obstacles before they arrived at the above correct recognition of the pathology of the disease. The spinal nature of the disease, first postulated by Heine in 1840 and later by Duchenne, was for a time called in question and the disease was described under the name of "essential infantile paralysis" (Barthez and Rilliet). Later, when Charcot, Joffroy and others had positively demonstrated changes in the spinal cord, it was supposed that poliomyelitis depended on a primary disease of the ganglion cells leading to atrophy, without any other disease of the spinal marrow (Charcot). This theory, which was based on the post-mortem findings in a case of poliomyelitis of some duration, suffered a severe shock when acute inflammatory changes in the spinal marrow were demonstrated (by Roger, Damaschino and Roth) in children who had died early in the disease; nevertheless, Charcot's conception of a primary disease of the ganglion cells has many adherents even at the present day (Risler and von Kahlden) and, up to a certain point, Lövegren). Most of the more recent investigators (Goldscheider, Redlich, Siemerling, Pierre Marie, Neurath), however, have expressed themselves in favor of an inflammatory nature of poliomyelitis and at the present time the discussion on this point—whether the disease is exclusively inflammatory—is practically confined to a controversy on the general problems of inflammation.

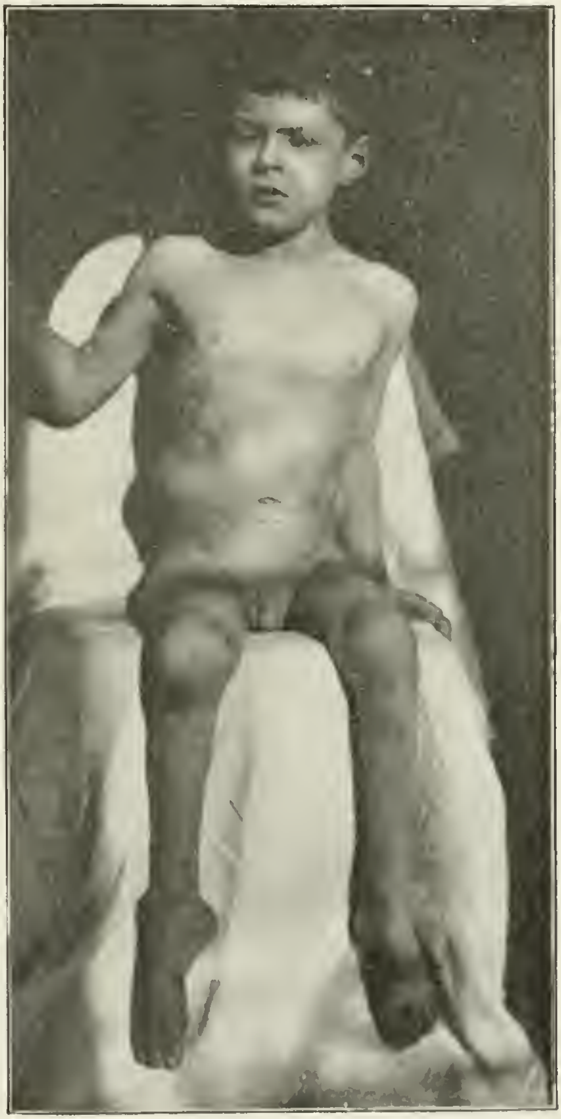
Fig. 45.



Diffuse cerebral sclerosis. Girl three years old. Gradual onset beginning in the third year of life. Spasm of the four extremities with emaciation and contractures. Apoplectiform seizures with disturbance of consciousness. Trismus and gritting of the teeth. Conjugate deviation; nystagmus; bulbar symptoms; atrophy of the optic nerve. No increase in the cerebral pressure; mentality but little affected. At the time the picture was taken the condition was stationary; possibly there was some improvement.

Recent cases of poliomyelitis exhibit post mortem all the signs of acute inflammation (hyperæmia, increase in the number of blood vessels, œdema, small cell infiltration, cloudy swelling of the ganglion cells, degeneration of the nervous elements and, ultimately, central softening) in the anterior horn. The inflammation is not limited to the gray matter but extends also to adjoining portions of the white substance. Extensive segments of the spinal cord are often involved in the inflammatory process; even the medulla oblongata may be attacked (Redlich). Localization of the lesions in the cervical and lumbar portions of the cord is characteristic of this form of inflammation of the spinal cord. In old cases the anterior horn on the diseased side is distinctly narrowed; the ganglion cells and fibres are atrophied or sometimes completely destroyed; and microscopically the vessels are often seen to be surrounded by the remains of inflammation.

Fig. 46.



Poliomyelitis. Boy seven years old. Atrophy and paralysis of both legs; the feet are fixed in a position of slight equinovarus. Paralysis of the abdominal muscles on the right side and protrusion of the abdominal contents, simulating hernia.

Numerous bacteriologic examinations have been made in poliomyelitis with the aid of lumbar puncture (Schultze, Bülow, Hanson, Harbitz, Dethloff and others). In most of the cases a diplococcus was found resembling that which causes cerebrospinal meningitis. Some French investigators have succeeded in artificially producing clinical pictures resembling poliomyelitis by injecting various bacterial cultures.

A number of external injuries were formerly included among the causes of poliomyelitis. Seeligmüller holds cold, infectious diseases, traumatism, muscular overexertion, dentition, psychic disturbances and heredity responsible for the development of poliomyelitis. But while all these may occasionally be contributory causes, the infectious diseases alone have any significance; for they frequently precede poliomyelitis so directly that the relation cannot be regarded as a mere accident. It must not be forgotten with regard to the significance of the history that the attending physician not infrequently mistakes the initial stage of poliomyelitis, when it is accompanied by fever, for some other disease and later, when he recognizes the extent of the poliomyelitis, is inclined to inform the parents that the paralysis is the result of the primary disease.

Aside from this form of postinfectious poliomyelitis, there is no doubt, however, that the disease in the great majority of cases occurs spontaneously and presents the character of a non-contagious infectious disease. This is shown by the massing of cases of acute myelitis during the summer months (July and August) (Barlow, Sinkler, Sachs, Zappert and Baumann), which is absolutely typical, and by the epidemic appearance of the disease which has been reported from various localities—America, Sweden, Norway, Switzerland, France, Italy, Germany and Austria. The Stockholm epidemic described by von Medin is particularly instructive from the fact that cerebral spinal and peripheral disturbances were observed at the same time.

Poliomyelitis is a disease of earliest childhood, most of the cases occurring in children who have not yet completed the second year. In half-grown individuals and adults the disease is much more rare and also exhibits certain differences in its course.

Poliomyelitis shows a preference for children who have previously been healthy. The initial symptoms are very variable. While it is true that prodromal symptoms, such as lassitude, anorexia and a low degree of fever are often overlooked, the onset of poliomyelitis is often so imperceptible that the children go to bed well the evening before, and wake up in the morning with signs of paralysis. More frequently the disease is ushered in by several days of fever of an uncertain type or by severe meningitic febrile states (the latter also when the disease is localized in the lumbar portion of the cord). The paralysis attains its maximum extent at the very beginning of the disease; gradual development of the paralysis by successful stages is the exception (Neurath and others). As soon as the initial maximum extent has been attained the paralysis in the extremities begins to diminish and ultimately is confined to a definite muscular region. This condition is permanent and is regarded as the recovery from the disease.

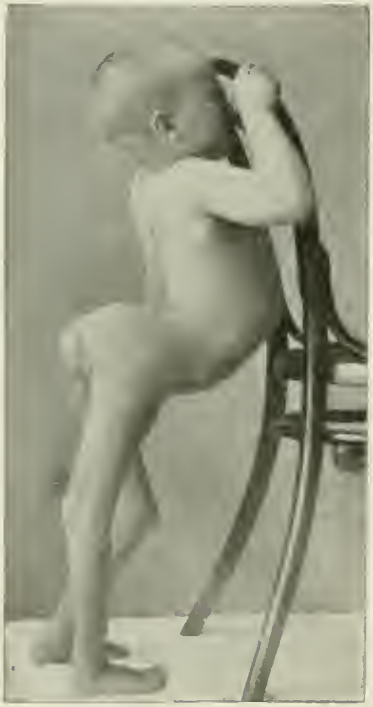
The characteristic sign of poliomyelitis is a flaccid, atonic muscular paralysis, accompanied by atrophy and loss of electric irritability. The paralysis is pronounced from the very onset, while atrophy and loss of electric irritability make their appearance within a few days. The muscles, shortly after the onset of the disease, exhibit loss of function but retain irritability wholly or in part and may be expected to recover. Various portions of the body are particularly liable to be attacked by the paralysis, a point on which there exist numerous statistics. The most frequent distribution is the two legs; next in frequency one leg, one arm, an arm and leg, either on the same or on opposite sides, and finally the muscles of the trunk and both arms.

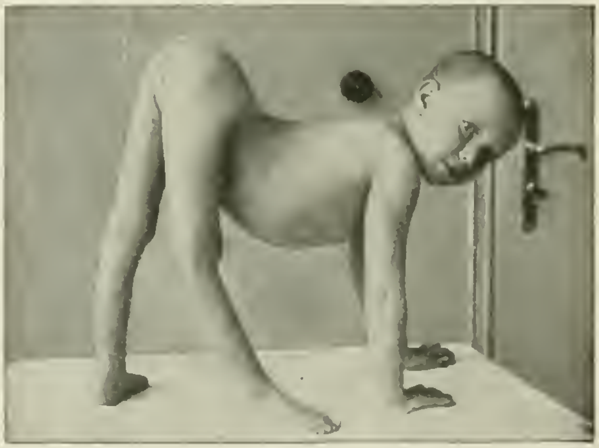
In the extremities the disease exhibits a remarkable predilection for certain muscles. In the arm, the deltoid, next the biceps, brachialis anticus, supinator longus (also the muscles of the upper plexus-type and the other shoulder muscles are chiefly attacked; paralysis of the triceps, of the muscles of the hand and complete paralysis of all the muscles of the arm are more rare. To sum up, therefore, the paralysis of poliomyelitis generally attacks the proximal portions of the arm. In the lower extremities the perineal muscles are very frequently affected. Next to these the tibialis anticus, the quadriceps, the gluteal muscles, the tibialis posticus and less frequently the muscles of the calf of the leg become paralyzed; the sartorius is never affected. Widespread flaccid paralysis of one or both legs is seen more frequently than the same condition in the arms. Paralysis of the muscles of the back with lordosis and disturbance of the gait, paralysis of the muscles of the neck causing dropping of the head, and paralysis of the abdominal muscles with unilateral or bilateral protrusion of the abdomen (Ibrahim) are rare, although I have personally seen all these forms. Associated paralysis of the muscles innervated by the cranial nerves (facial, hypoglossus and eye muscles) has also been observed a few times. The palsies may be combined in a variety of ways. Associated paralysis of several extremities (also in the form of spinal hemiplegia) is often seen in recent cases. As a rule, when different groups of muslces are affected, the paralysis subsides in one or more groups and the disease ultimately is confined to a much smaller territory than that of the original affection. Why certain muscle groups are affected more than others we do not know (Baumann); possibly the distribution depends on certain anatomical conditions such as the vascular supply of the individual muscle nuclei in the spinal cord.

The loss of function in poliomyelitis palsies may be very great. In the upper extremity the shoulder is completely paralyzed and rarely the other joints are also immovable. Poliomyelitis of the lower extremities produces flaccidity of all the joints involved (especially of the ankle-joint, "pendulum foot"); sometimes pes equinus with spasticity results owing to the preponderance of those muscle groups which are less severely involved. When the paralysis is unilateral the power of walking is very incompletely restored and may be permanently lost. Many of these unfortunate creatures have to resort to the most bizarre methods of locomotion ("hand walkers").

The tendon reflexes in the domain of the affected muscles are diminished or abolished, although a reflex that is absent at first may later reappear. The presence of a reflex, however, does not exclude the existence of spinal infantile palsy. For when the biceps, quadriceps, calf muscles (and the corresponding centres in the spinal cord) are intact, the triceps, patellar, and Achilles tendon reflexes may be preserved even if the other muscles of the same extremities are paralyzed. In fact, the reflexes in the leg may even be increased when the arm on the same side is paralyzed, probably because of a slight degeneration of the pyramidal tract due to extension of the primary inflammation to the region of the pyramidal tracts. The same explanation is probably applicable to the exaggeration of the Achilles tendon reflex when the knee jerk is absent, as sometimes occurs. The cutaneous nervous reflexes are present whenever the muscles concerned in their production are capable of reacting. Pain in the back or in the diseased extremity is common at the beginning of the disease (symptoms of spinal cord irritation) and later disappear (Duquennoy). Disturbances of the bladder are rare, at least in infantile poliomyelitis and, when present, usually represent an early symptom of involvement of the lower segment of the spinal cord. According to Oppenheim the possibility of isolated poliomyelitis affecting the muscles of the bladder and rectum (sudden occurrence of paralysis of the bladder and rectum) must be borne in mind. It must not be forgotten that sphincter paralysis in the initial stage may be due to hyperpyrexia and hebetude.

Figs. 47 and 48.





Poliomyelitis. Child seven years old. Severe paralysis of both legs, contracture at the hip-joint, marked lordosis of the lumbar portion of the spinal column ("hand-walker").

In addition to disturbances affecting the muscular apparatus, the poliomyelitis is accompanied by a number of changes in the osseous system and in the skin, which are generally designated trophic changes.

The skin of the affected extremities, particularly that of the legs, is cool to the touch, cyanotic and mottled like marble. There is a marked tendency to cutaneous diseases on the paralyzed side. While the eczematous eruptions which occur might be ascribed to the continual wetting incident to the use of electricity, other lesions resembling chilblains are probably due to vasomotor disturbances.

In early cases evidences of arrested development and a considerable degree of atrophy in the bones are quite frequently observed in the affected extremities, particularly the legs, and are readily demonstrated by means of the Röntgen rays. On the other hand, there are cases in which the diseased leg is longer than its fellow. Neurath offers for this peculiar condition the ingenious explanation that the child in such cases is probably rachitic and that the rachitis is less pronounced on the diseased side.

Flail joints, particularly in the shoulder, less frequently in the hip and knee, quite often result from paralysis of the muscles which support the joint and, it need hardly be said, considerably add to the already existing functional disability of the diseased extremity.

Scoliosis and lordosis are sometimes observed when the muscles of the back are paralyzed.

Among unusual anomolies, mention may be made of a tendency to the secretion of sweat (Higier) and hard œdema of the skin (scleroderma, Oppenheim).

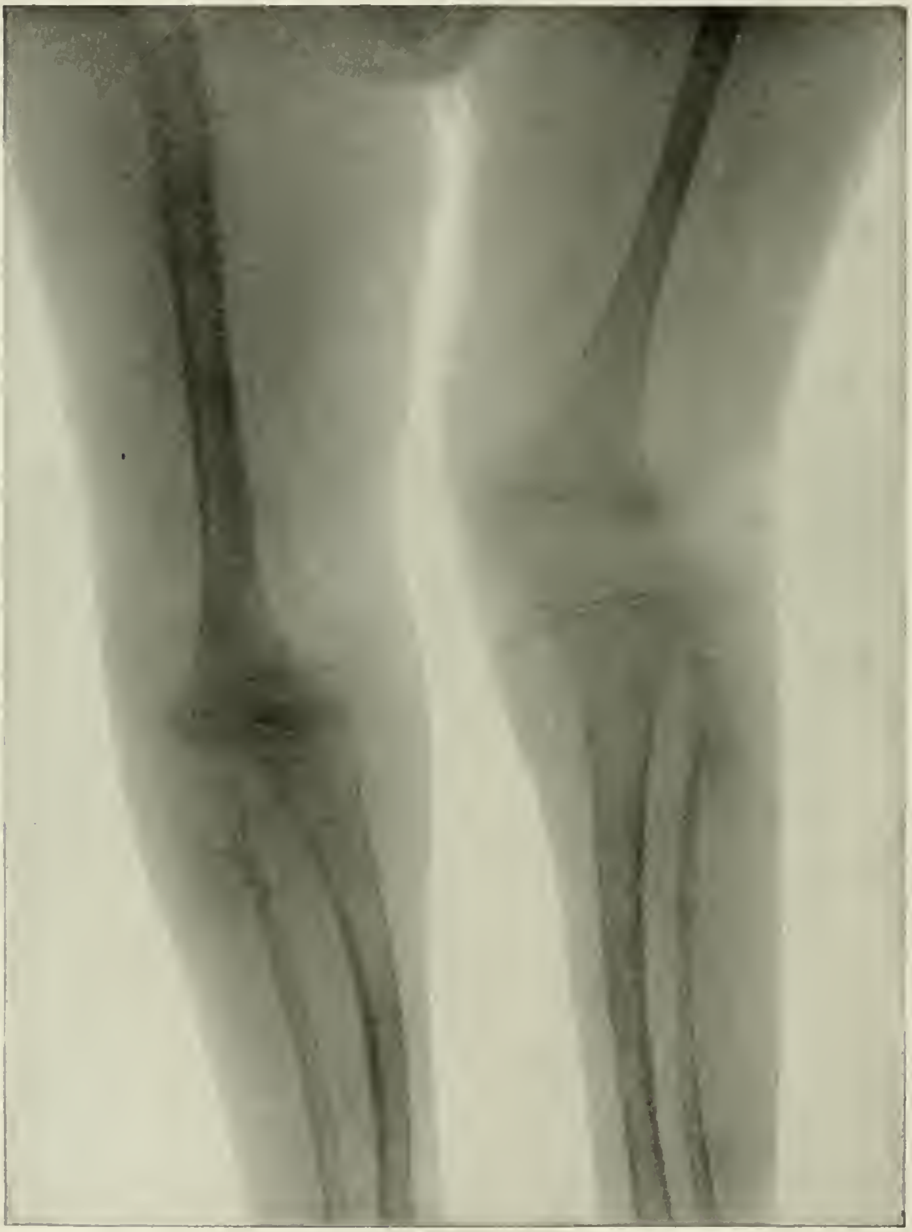
A few of these symptoms and their grouping in the various periods of the disease shows that in half way typical cases of myelitis three stages may be distinguished:

1. *Initial Stage.*—(*a*) This is often very brief and quite inconspicuous, without any striking symptoms (development of the paralysis over night); (*b*) or it may be accompanied by indefinite febrile symptoms which may last several days before the spinal paralysis manifests itself. (*c*) In other cases marked nervous symptoms are present from the beginning and point rather to a cerebral than to a spinal affection. It is possible that many of these cases terminate fatally with the picture of a meningitis, especially when the cervical portion of the cord is involved (Baumann), before the diagnosis of a spinal inflammation can be made. (*d*) Insidious onset without severe general symptoms, the disease reaching its highest point by gradual stages—in other words, a true subacute and chronic poliomyelitis—is extremely rare in children. In adults cases of this kind after traumatism or accident have a certain importance and probably rest on a different pathogenesis.

2. *Stage of Initial Paralysis.*—Immediately after the beginning of the disease a larger number of muscles are paralyzed than is the case later. Spinal hemiplegia, paralysis of both legs, paralysis of a leg and arm on opposite sides and, rarely, both arms, is observed. Many of the

muscles in a single extremity are often affected. Within a few days of the onset the preservation of normal electric irritability enables one to pick out the muscles that are capable of regeneration. Sometimes the varying degree of loss of function and the pronounced character of the tendon reflexes affords a clue as to which extremity or joint is

Fig. 49.



Poliomyelitis of both legs. Radiogram of both legs. On the left side the paralysis is much more pronounced than on the right. Distinct diminution in the size of the bones. (Especially of the femur on the left side.) The flail joint in the left knee is beautifully shown.

destined to remain permanently paralyzed. Sometimes pain and bladder disturbances are present. This stage usually lasts many months. At the beginning normal function is rapidly restored in some of the affected muscles, and even in those muscles which at first are persistently paralyzed considerable improvement often occurs after a time.

3. *Stage of Permanent Paralysis.*—In the end some muscle groups remain permanently paralyzed with loss of function and electric irritability. This constitutes the irreparable recovery from poliomyelitis with disability. Although even in this stage the affected members may retain a certain degree of function, this remnant,—aside from the effect of therapeutic measures,—depends on the compensatory action of other healthy muscle groups which is brought about by the growing intelligence. Atrophy of bones, flail joints, atrophic changes in the skin and deformities, are well marked in this stage.

Haushalter distinguishes 4 stages: (1) prodromal; (2) stage of the onset of paralysis; (3) stage of regeneration with permanent localization of the paralysis; (4) stage of muscular atrophy with deformity of the limbs.

The **prognosis** may be inferred from the above description. Although the well-marked changes in the spinal cord and the muscular atrophy to which they give rise are susceptible of very little improvement, a great reduction may nevertheless be expected to take place in the extent of the original paralysis. Even when the paralysis is stationary, the prognosis is not altogether unfavorable, since the important point to the patient is not whether certain muscle groups remain permanently atrophic but rather whether he will regain the power of performing certain necessary movements; and this power is often regained to a surprising degree by utilizing muscles that have not been affected by the paralysis.

The **diagnosis** of poliomyelitis may be difficult during the initial stage of the disease. The clinical picture is apt to suggest some general febrile disease, the beginning of an acute infectious disease, a meningitis or an encephalitis, and the sudden discovery of spinal palsies often comes as a great surprise.

The well-marked paralysis of the shoulder girdle is often difficult to distinguish from a birth palsy of the superior trunk of the brachial plexus, particularly in the absence of historical data. In a birth palsy the infraspinatus muscle is regularly involved, causing inward rotation of the arm, which is absent in poliomyelitis. Electric irritability is often unaffected or but slightly impaired in birth palsies. The diagnosis from inhibition palsies (Vierordt) of the arms may also present difficulties. Syphilitic pseudoparalysis is characterized by the general constitution of the child, the characteristic attitude of the hand, the osteochondritis which is usually present, and pain; *paralysie douloureuse (q.v.)*, by the well-defined onset of the paralysis with severe pain, the absence of atrophy or localization of the paralysis and, finally, by the rapid and favorable course. Rachitic pseudoparalysis may occasion diagnostic difficulties for a few days, the main points are, the less abrupt onset, the absence of atrophy, the uniform distribution of the muscular weakness in both legs and, finally, other signs of an existing rachitis. Oppenheim's myatonia, characterized by congenital flaccidity of the muscles of the leg with diminution or absence of electric irritability, abolition of the reflexes but presenting a favorable prognosis, is distinguished from poliomyelitis chiefly by the patient's age, the disproportion between the extensive functional weakness and the slight degree of atrophy and, finally, by the rapid results of electric treatment. Paralysis of the legs from spina bifida is characterized by the presence of local disease in the vertebral column (also in spina bifida occulta) and the existence of sensory and sphincter paralysis. Paralysis following hæmorrhage into the spinal cord cannot be distinguished from poliomyelitis in its ultimate results, as the same anatomical changes are finally produced in both cases. It is very probable that many cases of "congenital" poliomyelitis must be attributed to an intrapartum hemorrhage into the spinal cord; although cases of very early poliomyelitis have been observed (in a child 15 days old). Rapidly developing spinal palsy during whooping-cough may be attributed with equal justice to a hæmorrhage or to an acute inflammation, while a fall has been given as a possible causal factor; this should be accepted with caution as it may be the cause of a hæmorrhage just as well as an early symptom of poliomyelitis. The distinction between poliomyelitis and Hoffmann-Werding's spinal muscular atrophy is exceedingly difficult. Without a history and constant observation of the case the differential diagnosis is impossible, since in both conditions the lesion affects the spinal muscle centres. The rapidly progressive course of infantile spinal atrophy soon clears up any doubt in regard to the diagnosis. The diagnosis of Hoffmann's neural muscular atrophy is equally difficult; but it usually occurs in older children, is at first confined to the peroneal muscles, is bilateral, begins gradually and progresses slowly. Cerebral palsy acquired in early life and followed by arrested growth of one arm and under-development of the muscles, at the first glance presents some similarity to poliomyelitis, from which, however, it is distinguished by the persistence of electric irritability and of the reflexes, the fact that the paralysis is most marked in the hand, and by involvement of the facial muscles and the arm. The possibility of associated cerebrospinal palsies (simultaneous encephalitis and poliomyelitis) must however be kept in mind. Tumors of the spinal cord and spondylitis, even when they lead to atrophic palsies, are usually distinguished by their characteristic symptoms—pain, and involvement of the sphincters, and by the typical course of the disease. Multiple neuritis is rare in childhood and in case of doubt the weight of probability is in favor of poliomyelitis. The points in the differential diagnosis are, sensory disturbances, pain on pressure along the nerves, progress of the disease by successive attacks up to the point of its greatest development. If the polyneuritis leaves permanent palsies, the diagnosis is all the more difficult, as the palsies may possibly be due to a secondary spinal disease.

The **treatment** of poliomyelitis is quite different in the three stages of the disease.

If the physician is so fortunate as to see and recognize a case in the initial stage—which is very rare—he will have to content himself with general antipyretic and sedative measures (see treatment of encephalitis). Rest in bed, the application of ice bags to the vertebral column, free catharsis and, in older children, leeches would constitute the treatment. Lumbar puncture is to be recommended during these stages because, aside from the diagnostic results, it has a favorable effect on the inflammation by relieving pressure and possibly by removing bacterial poisons.

At the beginning of the stage of initial paralysis—the period when the physician usually sees the patient for the first time—it has long been customary to adopt a diaphoretic line of treatment, salicylates, aspirin, hot tea. It cannot be said that the improvement that occurs during this time can be attributed to the treatment. On the other hand, electricity and massage are of great value in this stage of poliomyelitis.

It is well to begin electric treatment about two weeks after the onset of the disease, keeping it up for many months, or even a year, three or four times a week. There are few organic palsies in the treatment of which patience and perseverance are so imperatively demanded as in poliomyelitis, and it is very probable that improvement in muscles that are completely paralyzed is hastened by electric treatment. The active pole, the cathode, of the galvanic current is used, and an attempt is made to produce a muscular contraction by interrupting the current. Especially devised electrodes are used for this purpose. Cathodal galvanization can also be effected by passing a current which produces twitching contractions over the paralyzed group of muscles. In this method of electric treatment the strength of the current is gradually increased and again diminished. If the muscles respond to the faradic current, the latter may be employed with the aid of a wandering electrode (for instance a faradic coil). If it is desired to produce a powerful effect, labile galvanofaradization, using both kinds of current, should be tried.

Next to electricity massage is of great importance in this stage of the disease, only however, when it is done by a practised hand and with accurate knowledge of the paralyzed muscles.

Warm baths are to be considered as an auxiliary measure and should precede passive movements and massage. Bathing cures with aerate thermal waters, warm saline or natural carbonated waters are useful during the summer, when the electric treatment is interrupted.

When the physician has become convinced that no improvement is to be expected by electricity or massage, orthopedic treatment is indicated. While this belongs to the stage of permanent paralysis, the ground may be prepared during the period of repair and the tendency to contracture may be combated by forbidding certain forced attitudes, and by the application of dressings and orthopedic apparatus. If contractures have already developed, various measures such as tenotomy, fixation of flail joints (arthrodesis), and various orthopedic apparatus are indicated in order to increase the functional power of the paralyzed extremities. The various apparatus devised by Hessing and Hoffa deservedly enjoy a wide popularity. A purely symptomatic and commendable surgical treatment consists in the transplantation of tendons after Nicoladoni. It is based on the principle of uniting the tendon of a paralyzed muscle to that of an adjoining healthy muscle. Only a part of the tendon of the healthy muscle is split off and attached to the paralyzed muscle so as not to destroy the function of the former. The close proximity of healthy and paralyzed muscles is indispensable for the application of this treatment and as this condition is satisfied in poliomyelitis more than in any other disease, it is here that the method has been chiefly developed. It is needless to say that the first requisite for the success of this operation is an accurate functional and electrical examination of the muscles to be operated upon, which alone enables the surgeon to map out his plan of procedure and this operation is quite difficult; after the muscles have been exposed, the difference in color and elasticity between the healthy and diseased muscles is of some importance. The best application of the method is found in partial palsies of the leg or of the arm. The results of this operation are very favorable (Lange, J. Wolff, Krause, Vulpius, and others). For the details of the technic, which has been carefully elaborated (for example, lengthening of the tendons with silk sutures, attachment of a transplanted tendon at the point of insertion of the paralyzed muscle) the reader is referred to Vulpius, Die Sehnenüberpflanzung, Leipzig, 1902.

VII. MYELITIS

In the present conception of poliomyelitis it is doubtful whether we are justified in describing myelitis in childhood as a nosologic entity. The difference, if there be any, is solely one of degree, depending on whether the inflammation involves the vascular domain of the entire cross-section of the spinal cord or is chiefly confined to the distribution of the anterior central arteries and therefore affects only the gray substance. It will be readily understood from the above-described symptomatology of poliomyelitis that an inflammatory affection of the spinal cord may begin as a myelitis and terminate as a poliomyelitis. There appears to be a difference between the spinal cord of children and that of adults as regards the manner in which the structure reacts to inflammatory irritation, and the seat of predilection of the disease also appears to vary at different ages. In the child inflammation of the spinal cord manifests itself chiefly as a poliomyelitis, in an adult as a myelitis. In the child the disease is found chiefly in the enlargements of the cord (cervical or lumbar portion); in the adult, chiefly in the dorsal portion. It may therefore be stated as an axiom that poliomyelitis represents the inflammation of the spinal cord characteristic of childhood and myelitis that of adult age.

The most important causes of myelitis are infectious diseases and intoxications. Traumatism and exposure to cold have lost much of the importance with which they were formerly credited. Bacteria have often been found in myelitic spinal cords, and the disease has also been produced experimentally. Syphilitic myelitis is discussed in section III. The pathology of myelitis consists in acute inflammation of the entire cross-section of the spinal cord, more often involving considerable portions of the structure. In cases which clinically suggest a transverse lesion, disseminated foci of inflammation are often found in the vicinity of the blood vessels. In old cases sclerotic changes, with proliferation of connective tissue, are seen.

The **symptoms** are determined by the seat of the inflammation. When this is in the thoracic portion of the cord—a frequent localization—there are paraplegia with exaggerated reflexes, anæsthesia of the legs, paralysis of the bladder and rectum, and bedsores. When the lesion is situated in the cervical portion, flaccid paralysis of the arms, with sensory disturbances are present in addition to the above symptoms. When the lesion is situated still higher up, oculopupillary symptoms and interference with respiration are to be expected. Myelitis of the lumbar portion gives rise to flaccid paralysis of the legs, with loss of sensation and paralysis of the bladder and rectum. Depending on the variety of paralysis present the reflexes are either increased or diminished, and electric irritability is normal or distributed. Twitching of the affected muscles ("spinal convulsions") particularly in response to external stimuli is not infrequent in myelitis. For the details of the symptoms which may occur in the various localizations the reader is referred to the tables in the sections on spina bifida and tumor of the spinal cord.

Myelitis begins with high fever and rapidly spreading spinal symptoms. The fever may persist for some time with renewed exacerbations (cystitis, bedsores) which may come on suddenly with chills. In rare cases the disease ends in complete recovery; or it may be arrested and permanent paraplegia result; or, finally, the child may die from one of the above-mentioned complications. For the pediatrist the most important cases are those in which a myelitic clinical picture actually develops the symptoms of a poliomyelitis.

The most important conditions in the differential diagnosis are tumor of the spinal cord and spondylitis.

Abscess of the spinal cord is extremely rare. It results from traumatism or, like brain abscess, may be metastatic. The favorite seats are in the upper portion of the spinal cord with a predilection for the gray substance. The symptoms are chiefly those of acute inflammation of the spinal cord, with intermittent fever. Such symptoms can be ascribed to an abscess only when a cause for the suppuration can be demonstrated (Schlesinger).

VIII. LANDRY'S PARALYSIS (ACUTE ASCENDING PARALYSIS)

In 1859 Landry described a clinical picture which consists in flaccid paralysis of the skeletal muscles, beginning in the lower extremities and rapidly ascending, and terminates fatally in a short time through involvement of the bulbar nerves and the respiratory centre. The pathologic findings in Landry's cases were negative.

Since his time the disease has been studied by many clinicians and pathologists, among whom Westphal, Bernhardt, Kahler, Pick, Wappenschmidt, etc., have contributed greatly to our knowledge of it. It is observed in individuals of every period of life. Cases occurring in children have been described among others by Liegard (2½ years old), Kahler, Pick, Heubner, Soltmann, Gru, Rumpf, etc. On the whole, however, the disease is distinctly rare.

The first **symptom** is paralysis of the toes and feet, sometimes accompanied by dragging pains and paræsthesia. Within a few hours the entire leg becomes paralyzed. By the end of several more hours, or at most a day, paralysis is complete in both legs. After a short interval the muscles of the trunk, back and thorax are attacked in turn, and within a short time the arms also are paralyzed, the paralysis beginning in the shoulders and fingers. But the dreadful disease continues to spread; alarming interference with deglutition, speech and respiration develops, and the unfortunate patient is finally robbed of all power of movement except the movements of the face and eyes. Death occurs from asphyxia, and consciousness is retained to the end. Aside from paræsthesia and sensations in the extremities before the appearance of the paralysis, pain is usually absent. The cutaneous and tendon reflexes are absent, electric irritability disappears or the reactions of degeneration are present, feeble stimuli being often sufficient to produce severe contractions. The sphincters escape, and there are no trophic disturbances. If the disease begins with bulbar symptoms, death occurs before the paralysis becomes universal. Fever is not present, as a rule, but has been described in children (Soltmann).

The average duration of the disease is from 1 to 1½ weeks. Most of the cases end fatally, although arrest of the disease has been observed at every stage, even that of bulbar involvement.

Our knowledge of the pathology, anatomy and pathogenesis of this disease is still incomplete. In the cases which have been examined so far the findings were either negative or those of acute inflammation of the spinal cord (poliomyelitis acutissima) and of the medulla oblongata, neuritis of the roots of the spinal nerves, or, finally, acute polyneuritis. With the newer methods of examination negative findings are becoming more and more rare. On the strength of these pathologic findings and the corresponding clinical symptoms we distinguish a medullary, bulbar, and neuritic form of Landry's paralysis (Leyden-Goldscheider). But the identity of all the forms of this disease is now no longer questioned, and the theory of an acute intoxication is generally accepted, the differences in the clinical picture being explained by the point of attack, the severity of the intoxication, and the duration of the disease (Oppenheim, Remak). It is difficult to determine whether the disease is localized in the spinal cord or in the peripheral nerves, and Raymond's designation *cellulo-neurite aiguë antérieure* therefore has much to recommend it.

The nature of the intoxication that is responsible for Landry's paralysis is unknown. The disease has been observed after anthrax, diphtheria, influenza, typhoid fever and gonorrhœa, and the corresponding microörganisms have been demonstrated in the pathologic preparations.

The only conditions with which the disease could be confounded are some spinal affections (spinal infantile muscular atrophy and poliomyelitis) and polyneuritis, and the diagnosis is speedily settled by the rapid course of the disease. The prognosis is unfavorable. Mercury and ergotin have been recommended. Counterirritation with the actual cautery has also been recommended, but whether such an heroic measure is justifiable in the case of a patient's suffering from such a pitiable disease must be left to the physician's own feelings.

SECTION VI.

NEOPLASMS OF THE CENTRAL NERVOUS SYSTEM

(MULTIPLE CEREBRAL AND SPINAL SCLEROSIS)

The study of neoplasms of the central nervous system has taught us that the symptomatology is determined more by the seat of the tumor than by the nature of the neoplasm. Text books, therefore, with few exceptions (Henoch's Lehrbuch) discuss genuine neoplasms and the granulation tumors together.

The same plan will be adopted in this work although it cannot be denied that the signs of brain tubercle in children are sufficiently characteristic to justify a separate classification.

Brain tumors are very frequent in childhood. Gowers calculates that one-third of all the cases that have been analyzed occurred during the first two decades of life. Brain tubercle greatly preponderates over other tumors (out of 62 cases of brain tumors, examined post mortem in the Karolinen-Kinderspital in Vienna, 53 were tuberculous). The accompanying table by Allen Starr gives a good idea of the character, frequency and localization of brain tumors in childhood and renders a detailed discussion of the subject superfluous. The frequency of cerebellar tumors in childhood may however be emphasized.

BRAIN TUMORS IN CHILDREN AND ADULTS

The first column contains tumors in childhood; the second tumors in adults.

	Tuberculous		Gliomatous		Sarcomatous		Glio-sarcomatous		Cysts		Carcinomatous		Gummatous		Undetermined		Total	
I. Cerebral axis																		
1. Basal ganglia and lateral ventricles	14	3	3	9	5	8	.	.	1	1	1	2	.	1	3	5	27	31
2. Corpora quadrigemina and cerebral peduncles	16	1	1	2	3	2	.	5	..	.	.	1	.	..	1	7	21	11
3. Pons	19	11	10	..	5	1	2	1	1	.	.	2	.	3	1	..	38	17
4. Medulla oblongata	2	..	..	1	..	..	.	.	1	.	2	..	.	..	1	..	6	2
5. Base	..	3	..	2	1	3	1	1	1	.	1	..	.	..	4	1	8	9
6. Fourth ventricle	1	..	1	..	1	1	.	.	..	.	1	2	.	..	1	1	5	4
II. Cerebellum	47	8	1	3	10	13	1	6	9	.	3	..	.	..	11	10	96	45
III. Multiple tumors	34	4	15	2	3	5	.	.	2	.	.	2	1	3	3	..	43	17
IV. Cerebral cortex	13	9	..	19	1	46	.	8	..	1	1	19	.	13	1	12	21	127
V. Centrum ovale	6	2	1	11	5	7	1	4	15	.	1	3	1	..	5	14	35	51
	152	41	37	54	34	86	5	25	30	2	10	33	2	20	30	41	300	300

Even infants are subject to the disease (Demme's case in an infant three days old; infant five months old in the Karolinen-Kinderspital with brain tubercle). Aside from congenital tumors (angiomata), the most common are tubercles, but sarcomata also occur during the first year of life (ten-months-old infant in the Karolinen-Kinderspital). The greatest tendency to tuberculosis of the brain exists during the first years of life. The male sex furnishes the largest contingent of brain tumors (according to Gowers 440 men and 220 women). This disproportion is attributed to the fact that men are more exposed to head injuries, which are supposed to play an etiologic rule in many forms of tumor.

Pathologic Anatomy.—In the majority of cases brain *tubercle* is multiple (in 35 out of 53 cases in the Karolinen-Kinderspital). The seats of predilection are the cerebellum (24 out of 53 in the Kinderspital), the basal ganglia, the pons, the corpora quadrigemina, the cerebral hemispheres and, rarely, the medulla oblongata. The tumors are spherical, with a poor blood supply, necrotic at the centre, caseous, and surrounded by a layer of vascular granulation tissue which contains giant cells and tubercle bacilli. Children who die of brain tuberculosis frequently have a terminal tuberculous meningitis and always present other signs of tuberculosis, particularly in the glandular system. *Glioma* is a tumor consisting of neurogliar tissue which occurs only in the central nervous system and is usually quite different from sarcoma (Strube). Gliomata are found quite as often in the cerebrum as in the cerebellum, and frequently in the pons. In contradistinction to tubercle, glioma is apt to present itself in the form of a flat proliferation extending laterally rather than as a spherical tumor; in fact the infiltrating tumor may be so intimately mingled with the brain substance as to produce an apparent hypertrophy of certain portions of the brain. In color and general appearance glioma resembles the tissues of the brain, but the neoplasm is hard, more vascular and more reddish. The composition is sometimes more fibrous (fibroglioma), or mucoid (myxoglioma). Hæmorrhagic and cystic softening is not infrequently seen in glioma. *Sarcoma* shows a predilection for the cerebellum, but also occurs quite frequently in the cerebral hemispheres, especially when the growth starts in the calvarium or the dura mater and the brain is involved secondarily. When a sarcoma of the dura mater breaks through it is sometimes called a *fungus duræ matris*. Sarcoma is a typical malignant neoplasm and grows rapidly, compressing the cerebral mass and producing necrosis; it also breaks through from one tissue to another without respecting tissue boundaries. According to the nature of the supporting tissue, we distinguish fibro- myxo- and, particularly, vascular angio-sarcoma. Necrosis also occurs in the interior of sarcomatous tumors. Cavernous angiomata, which are congenital and grow rapidly after birth, may involve not only the surface of the brain but also the meninges, the bones of the skull and even the external coverings (Kalischer). *Carcinoma*, usually soft and vascular, sometimes occurring as a primary cerebral or dural tumor, *psammoma* (fibrous neoplasms of the pineal gland mixed with brain sand), *cholesteatoma* (a pearly gray epithelial tumor) and *adenoma of the hypophysis* are rare forms of tumor in childhood. *Parasitic cysts* (cysticercus, echinococcus) are more important because they are as frequent in children as in adults. *Cysticercus* usually forms small multiple vesicles on the surface of the brain and in the ventricles. Within the vesicles the head of the worm is seen as a black point (with a microscope the sucking organs can be recognized). As sequelæ localized inflammations of the brain and of the meninges are observed. *Echinococcus* of the brain is much more rare and leads to the formation of a much smaller number of vesicles, which are larger in size than those of cysticercus. Parasytic cysts may undergo calcification with complete recovery.

Secondary changes, due to the rapid growth of the tumor, are almost always seen in post-mortem examinations of brain tumors, particularly internal hydrocephalus, flattening of the cerebral convolutions and erosion of the cranial bones.

Symptoms.—The symptoms of brain tumor are subdivided into those which are produced by the increased pressure in the brain and those which are due to the seat of the neoplasm. The most important pressure symptoms are headache, vomiting, vertigo, choked disc, slowing of the pulse and convulsions. Headache, one of the earliest signs of brain tumor, may be persistent or occur paroxysmally, and in the latter case particularly may be excessively violent. When the pain is localized in the occiput and radiates into the neck, a cautious diagnosis of neoplasm in the posterior cranial fossa may be made. Marked variation in the intensity of the headache is regarded by Allen Starr as a sign of great vascularity of the tumor and therefore points rather to glioma or sarcoma than to tubercle. Localized pain, elicited by percussing the skull, sometimes enables the examiner to locate the seat of a neoplasm. Vomiting usually coincides with the period of greatest intensity of the headache and, in general, is more marked during the beginning of the disease, becoming rarer if the duration is protracted. It comes on suddenly—"projectile vomiting"—and does not always bring the relief that follows gastric vomiting.

Owing to the occurrence of choked disc and the impairment of vision which it causes, older children with brain tumors are often seen first by the oculist. Choked disc is an early characteristic symptom, particularly when the tumor is situated in the cerebellum, crus cerebri or the base of the brain; in fact, optic neuritis with atrophy may occur in these cases before other symptoms of tumor are present. On the other hand there are brain tumors, particularly multiple tubercles, in which choked disc occurs late or not at all and causes only slight discomfort. Oculists now-a-days make a sharp distinction between choked disc, accompanied by marked œdema at the entrance of the optic nerve, and inflammation of the optic nerve or optic neuritis. The former is a concomitant symptom of brain tumor only; the latter is seen with every kind of intracranial inflammatory process. Ophthalmologists are divided on the question whether choked disc is the result of the increase in intracerebral pressure or of a secondary inflammatory œdema; the majority are in favor of the former view. Unilateral choked disc and hemianopsia point to a disease focus in the chiasms or in one of the optic nerves. Persistent or paroxysmal vertigo may be a general symptom of brain tumor or the result of disease of the cerebellum or of one of the crura cerebelli, or it may accompany palsies of the ocular muscles. Slowing of the pulse is particularly frequent at the height of an attack of headache, especially in diseases of the posterior cranial fossa. Marked variations in the pulse are frequently observed as the patient changes from the recumbent to the erect position, particularly if the change is sudden. During the terminal stage slowing of the pulse (irritation of the vagus) is replaced by acceleration (paralysis of the vagus). Yawning, sobbing and anomalies of the respiration are other concomitants of brain tumor, especially in the advanced stage of the disease. Psychic changes also, particularly depression, disinclination to play, anorexia or even persistent hebetude belong to the picture of brain tumor. Disturbance of speech is not rare and is not necessarily a focal symptom. Convulsions, both general and cortical, are not at all infrequent, especially in brain tubercle in children. Many cases of this disease are first recognized by the sudden occurrence of convulsions; and paralysis of the extremities, accompanied by convulsions, is not infrequently the first symptom. General convulsions are also quite common during the terminal stage of brain tumor in children and, in cases of brain tubercle, may be attributed to a secondary meningitis. Cortical (Jacksonian) convulsions are observed in brain tumor at the beginning of the disease, and usually indicate that the tumor is on the surface of the brain. They consist in localized twitching of the face, or in one arm or leg, at first without disturbance of consciousness, the latter occurring only when the convulsions attack the other side of the body also. Loosening of the cranial sutures and enlargement of the circumference from secondary hydrocephalus are frequent symptoms of brain tumor in early childhood. Percussion of the skull, particularly of the frontal and parietal bones, gives a ringing sound similar to the cracked-pot sound heard over a pulmonary cavity.

Focal Symptoms.—These result in part from destruction of a portion of the brain, and partly from pressure of the tumor on adjacent or remote portions of the cerebrum (direct focal symptoms, pressure symptoms, and remote pressure symptoms). The symptoms may be those of irritation or of paralysis. Irritative symptoms are particularly frequent in the facial muscles and in the extremities, and consist in tremor, choreic and athetoid movements, cortical epilepsy and hemiplegic convulsions. They are usually the forerunners of the actual palsy. Irritative symptoms in the organs of special sense may also precede loss of the respective function.

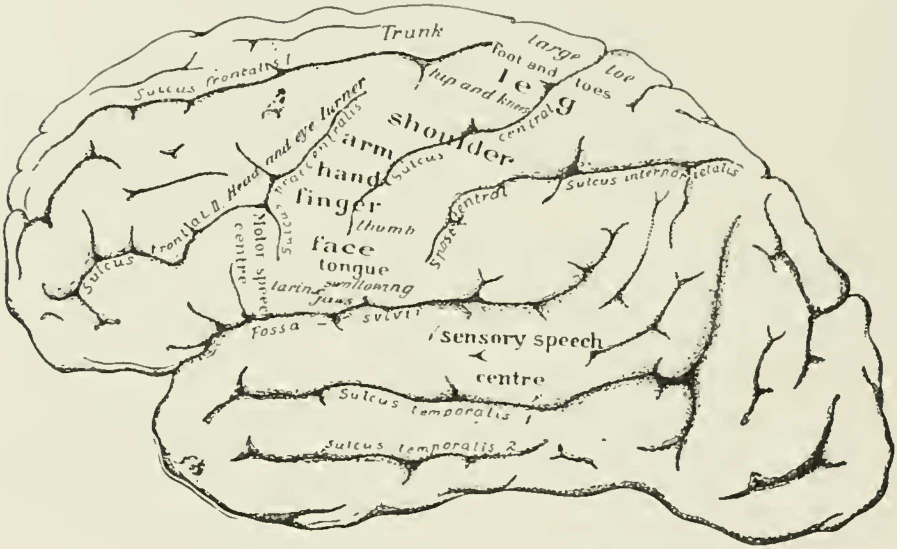
It is obvious that focal symptoms may be produced by circumscribed disease of the brain other than a neoplasm; hence the following paragraphs apply to all forms of cerebral disease. We shall not attempt to give a detailed description of focal symptoms and refer the reader for that purpose to the books of Bruns, Gowers, Oppenheim and others.

1. *Central Convolutions* (motor area).—Hemiplegia of one arm, one leg, or one-half of the face; the paralysis can only spread from one cortical field to another, as shown in Fig. 50. Thus, for example, paralysis affecting one leg cannot extend to the face without involving the arm. Frequent onset with cortical epilepsy; not rarely paræsthesia in the extremities, especially before the convulsive attacks. Accurate focal diagnoses can be made by noting the progress as indicated by each successive attack or the gradual extension of the paralysis. Ocular symptoms (conjugate deviation, strabismus) are very common in irritative conditions affecting the surface of the brain.

2. *Frontal Lobe.*—Motor aphasia in diseases of the posterior portion of the third frontal convolution of the left hemisphere (Broca's region); hence diseases of the central convolutions on the left side are practically always accompanied by disturbances of the speech, either partial or total aphasia. Other disturbances characteristic of disease of the frontal lobe (diminished intelligence, a tendency to crack jokes—so-called frontal ataxia) cannot very well be recognized in the child. Hemiplegia from pressure on the motor tracts is sometimes a remote symptom.

3. *Temporal Lobes.*—Sensory aphasia from lesion of the first convolution of the left temporal lobe (Wernicke); auditory disturbances which, however, are marked only in bilateral lesions; possibly disturbances of smell and taste (centres in the uncus gyri hippocampi?) Among remote effects, symptoms of irritation of the motor region; hemianopsia. Marked disturbances of all the functions of speech are found in lesions affecting the island of Reil.

Fig. 50.



Motor regions on the convex surface of the cerebral cortex in man. The regions whose functions are still in doubt are indicated in small print to distinguish them from the well-established motor regions.

4. *Occipital Lobe.*—Crossed homonymous hemianopsia without hemianopic pupillary reaction; sometimes symptoms of optic nerve irritation (flashes of light). In rare cases optic aphasia and alexia; and, finally, psychic blindness, usually in bilateral lesions.

5. *Basal Ganglia* (corpus striatum, lenticular nucleus, optic thalamus).—Sometimes without symptoms; usually the effects of remote pressure on the internal capsule with gradual development of a simple hemiplegia, often accompanied by irritative symptoms in the extremities. In tumors of the *optic thalamus* contralateral athetosis, disturbances

of sensation and unilateral facial paralysis should be mentioned. Tumors in the centrum ovale may be present for a long time without focal symptoms; later, there may be symptoms of irritation or paralysis, the nature of which will depend on the nearest motor or sensory tract.

6. *Corpora Quadrigemina.*—Ocular palsies (particularly of the external oculomotor and trochlear nerves) of unequal intensity on the two sides, which are not involved at the same time; often associated. Cerebellar ataxia; often tremor of the arms; unilateral deafness; possibly heminanopsia.

7. *Crus Cerebri.*—Alternating hemiplegia, *i.e.*, paralysis of the oculomotor on the diseased side and of the facial, extremities and hypoglossus on the opposite side, often with tremor of the extremities (*syndrome de Benedikt*). Later the oculomotor on the opposite side also becomes involved.

8. *Pons.*—Alternating hemiplegia as follows: facial, abducens, trifacial, either on both sides or only on the side of the tumor; crossed paralysis of the extremities; sometimes also involvement of the cranial nerve centres in the medulla (pressure). Often associated ocular palsies. Sometimes the cranial palsies first spread to the other side (of the abducens for instance) and the extremities are not involved until later; rarely disease of the auditory nerve.

9. *Medulla Oblongata Including the Fourth Ventricle.*—Symptoms referable to the auditory, glossopharyngeal, pneumogastric, hypoglossus, medullary nucleus of the spinal accessory; also deafness, paralysis of the muscles of deglutition; difficult speech; aphonia; atrophy of the tongue; disturbances of the cardiac and respiratory functions; crossed paralysis of the extremities. Rapid extension, involvement of the opposite side; simultaneous involvement of several cranial nerves (in basal lesions the nerves are attacked in succession). Tumors in the fourth ventricle, such as cysticerci floating free in the cavity, give rise to very inconstant symptoms without any local signs of loss of function.

10. *Cerebellum.*—Cerebellar ataxia; vertigo; rapid appearance of severe symptoms of cerebral pressure, particularly choked disc, headache, vomiting, stiffness in the muscles of the neck. Remote pressure symptoms referable to the quadrigemina, the pons and the medulla oblongata, with the above-mentioned symptoms which follow the ataxia. If these remote symptoms are distinctly present only on one side of the body, it may be possible to locate the tumor in one or the other half of the cerebellum.

11. *Base of the Brain.*—When the tumor begins in the bones or other structures at the base of the skull, rupture into the eyes, nose, or pharynx often occurs (exophthalmus). Very violent pain; early involvement of the eyes; often unilateral, associated palsies of several cranial nerves. The tumor can sometimes be seen in the X-ray photograph

(Oppenheim, Schüller). *Anterior fossa of the skull:* Aside from the above-mentioned general symptoms there are few local symptoms, which correspond to that portion of the cortex which is injured by the pressure. *Middle fossa of the skull:* Disturbances of the optic nerves, of the visual field (bitemporal hemianopsia), blindness. An amaurosis may occur before the development of choked disc or atrophy of the optic nerve. Later, ocular palsies (ptosis); symptoms of trifacial irritation (neuralgia, facial anæsthesia, atrophic paralysis of the muscles of the jaws, neuroparalytic keratitis). Tumors of the *hypophysis* produce the same symptoms and, in addition, obesity and hypoplasia of the genital organs (see also acromegaly). *Posterior fossa of the skull:* Pressure symptoms referable to the cerebrum, the pons and the medulla oblongata. The occurrence of ataxia is preceded by paralysis of the cranial nerves, which develops more slowly than when the lesion is situated within the medulla. Often association of facial and auditory paralysis, especially in primary tumors of the auditory nerve (neurofibromatosis, Oppenheim).

The following definitions are given to explain some of the above-mentioned symptoms:

Motor aphasia is inability to form words, although the power of understanding the words is preserved; in children it appears that when the speech centre on the left side has been destroyed, a new centre may be developed on the right to take its place.

In *sensory aphasia* (word deafness) the patient can speak and hear spoken words, but does not understand their meaning. He is approximately in the position of "a foreigner who does not understand our language."

In *optic aphasia* the patient is unable to name an object that is shown to him, although he is perfectly acquainted with its nature and in conversation speaks of it by its correct name.

Agraphia is the loss of ability to write, although the motor function of the arms is not affected. *Alexia* is inability to read in individuals with normal vision, who have been able to read all their lives.

Psychic blindness manifests itself in inability to associate with visual impression of an object a proper conception of its use, size, distance, etc.

In *bilateral homonymous hemianopsia* one half of the retina of both eyes on the side corresponding to a lesion of the optic tract or of the occipital lobe is insensitive, hence the opposite halves of the visual field are not seen.

In *bitemporal hemianopsia* both temporal fields are wanting owing to insensitiveness of the two median retinal halves (occurs in diseases of the chiasm).

Correct interpretation of the pressure symptoms and an attempt to determine the seat of the neoplasm from the focal symptoms that are present usually exhaust the possibilities of accurate diagnosis. To determine the *character* of the neoplasm is much more difficult and can usually be done only by other concomitant symptoms.

In children, the most important diagnosis is that of *tubercle,* and we shall therefore add a few remarks about the various forms which it may assume. As a rule the disease occurs in children who have previously shown scrofulotuberculous symptoms (glandular enlargement, suppuration from middle ear, diseases of the eyes, cutaneous tuberculides or bone caries). The brain symptoms may come on gradually with headache, vomiting, peevishness, so that tuberculous meningitis appears more likely than brain tumor. If the eyegrounds are examined at this stage, the condition may be explained by the finding of a choked disc; if this is absent, as is often the case, especially in multiple tubercles of the cerebrum, the absence of further signs of meningitis, the occurrence of localized palsies or symptoms of irritation, and the fact that the general brain symptoms remain constant for some time, will awaken the suspicion of a brain tumor. In other cases the indefinite initial stage is interrupted by cortical or unilateral convulsions, which are often followed immediately by paralysis. Gradually developing unilateral palsies not infrequently constitute the only symptom of a brain tumor and, if other pressure symptoms are indistinct, may for a time cause confusion with infantile cerebral palsy. Instead of a simple spastic hemiplegia, tremor, chorea or athetoid movements—without any symptom of tumor—sometimes give the first intimation of the presence of brain tubercle. Indeed, the picture of bilateral chorea may be simulated by tuberculosis of the brain. Finally, brain tubercle in the child quite often produces no symptoms whatever; the patient may present the picture of a possibly not quite typical meningitis and at the autopsy a solitary tubercle is found in the brain. It must of course not be inferred from the present description of the more obscure forms of brain tubercle, that typical cases of the disease, that is, cases with distinctly localizable focal symptoms and characteristic signs of brain pressure, are rare in childhood. On the contrary, tubercle situated in the pons, in the corpora quadrigemina, in the cerebral peduncles and in the cerebellum quite frequently furnish instructive examples for the focal diagnosis of a neoplasm and permit the observer to follow the slow growth of the tumor quite distinctly by the clinical signs.

The symptomatology of *glioma* and *sarcoma* does not differ materially in the child from that of the same conditions in the adult. The presence, of such a neoplasm should be suspected when in a previously healthy vigorous child symptoms of tumor develop rapidly. The height of the disease is reached in a shorter time than is usually the case with tubercle. In cases of neoplasms situated in the cerebellum and in the posterior cranial fossa, which are so frequent, both the general, and the focal symptoms early assume great importance.

Cysticercus gives rise to remarkably few local symptoms; not even pressure symptoms are always pronounced. Some of the many different symptoms of these tumors are headache, vomiting, general or localized convulsions, muscular spasms (often associated with twitching in the shoulder and in the face), mental confusion, depression, as well as typical cerebellar and bulbar symptoms. Choked disc is usually absent. In cases of cysts floating free in the ventricles alternation between the picture of grave disease and good health are not rare, so much so that the patients are regarded as hysterical or neurasthenic until, to the surprise and discomfiture of the physician, severe permanent symptoms (blindness) or death suddenly occur.

Tumors of the *hypophysis,* mostly in the form of adenoma, rarely psammoma or sarcoma, have but little significance in children. Adenoma of this organ, which develops in later life, gives rise to a peculiar clinical picture, that of *acromegaly,* the cardinal symptoms of which are increase in the size of the hands, feet and bones of the face; thickening of the skin, disinclination to work, apathy, more rarely bulimia, polydipsia, paræsthesia and pain. The signs of tumor are not necessarily present. As a rule the thymus gland persists. The course is slow and chronic, and the disease is not directly fatal. The symptoms of this condition are due not to the presence of the neoplasm but to the disturbance of the internal secretion of the hypophysis; in fact, the presence of the hypophyseal tumor in acromegaly is now regarded rather as a secondary phenomenon of the disease than as its cause.

The course of brain tumor in children is not less grave than in adults. It is true that the course is not rapid, in the case of brain tubercle particularly, which is so frequent; so much so in fact, that the slow development of the disease is a valuable diagnostic point between tubercle on the one hand, and glioma and sarcoma on the other. Death in cases of tubercle is very frequently due, not to the effects of the tumor itself, but to a tuberculous meningitis or a general miliary tuberculosis. Rapidly growing tumors of the posterior cranial fossa, particularly sarcoma and glioma, may cause sudden death. In general, however, a duration of several years is not uncommon in cases of brain tumor in childhood, particularly cases of tubercle; toward the end the symptoms assume a violent character and death occurs within a few days.

Whether recovery in cases of brain tumor is possible is exceedingly doubtful (syphilitic diseases excepted). In the case of tubercle there is of course a possibility of calcification taking place but, judging from the autopsy findings at our disposal, it appears to be a very rare event. Cysticercus, on the other hand, undoubtedly does calcify, with subsequent recovery. Of clinical examples of recovery from a tumor there is no lack and I have personally seen such cases. They must, however, be accepted with great caution; for it must be remembered that long periods of latency are among the possibilities in cases of brain tumor (tubercle) and, on the other hand, it is very probable that circumscribed encephalitis, in which recovery is undoubtedly possible, may present a clinical picture in every respect similar to that of tumor (see encephalitis).

The **differential diagnosis** has but a limited field in childhood. Neoplasms occurring in children suffering from hereditary syphilis ought not to be classified as tumors, because they are rarely uncomplicated and it is practically impossible to differentiate clinically between gumma, encephalomalacia and a circumscribed meningitis. Cases of tumor with hemiplegia and paraplegia are often mistaken for cerebral infantile palsy. The diagnosis rests on the ophthalmoscopic findings, the history (slowly acquired affections are not cerebral infantile palsies), and the course of the disease, which is progressive in tumor and retrogressive in infantile palsy. It has already been mentioned that brain tubercle is not infrequently confounded with meningitis, especially the hæmorrhagic form of the disease. It has also been pointed out that the first symptoms of a tumor, and the variable picture produced by cysticercus floating free in the ventricles may simulate hysteria or neurasthenia. Headache, vomiting, and signs of general malnutrition quite frequently suggest the diagnosis of gastric trouble, and the presence of a tumor is recognized only by the discovery of choked disc, by the progressive course, and by the focal symptoms which sometimes make their appearance. For the differential diagnosis between brain abscess and encephalitis, see the respective sections.

Much information of value for the diagnosis may be expected from *radiography* and *lumbar puncture*. A good X-ray picture will sometimes reveal the presence of a basal tumor. On the whole, however, the method is not often applicable. Lumbar puncture is of value chiefly when the findings are negative, *i.e.*, when the fluid obtained is under high pressure, clear like water, free from bacteria and coagulable, it points to meningitis. Very rarely particles of tumor are found in the fluid.

.The **treatment** of brain tumor is still as hopeless as it ever was. Attempts at a casual therapeusis by operative removal of the tumor, made in recent years, while they reveal a high degree of acumen as well as accuracy in diagnosis, promise but little success in the case of children. This is partly because children tolerate the very bloody operation badly and partly—and this is more important—because the seat and variety of the tumors that are most common in childhood offer less favorable chances for successful operative removal. Tumors in the cerebellum, the basal ganglia and the pons, which are more frequent than any others, must still be regarded as practically inoperable, and brain tubercle, on the other hand, is frequently multiple and is the expression of a constitutional disease so that the removal of one diseased spot does not insure the patient's recovery. Thus, among a very large number of cases of brain tumors that have come under my observation in the course of years, I can call to mind only a single one in which I could advise operation. This was in a vigorous boy without any tuberculous taint, in whom a progressive monoparesis of one arm had developed along with characteristic signs of tumor; but the parents could not bring themselves to consent to the operation. According to the local indications for operative intervention set up by Bruns tumors in the following situations may be regarded as operable: (1) the central convolutions; (2) the speech regions; (3) the frontal lobes; (4) the occipital lobes; (5) the temporal lobes. These indications will have to be followed in the few operable cases that occur in childhood.

If there is no indication for the operable removal of a brain tumor, a palliative operation still remains to be considered and should be employed symptomatically when the paroxysms of pain are very severe or there is beginning atrophy of the optic nerve. The procedures are lumbar puncture and trephining of the skull, of which the former is the more important from the pediatrist's standpoint. Unfortunately lumbar puncture is not without danger in cases of brain tumor, because the sudden removal of pressure in the brain may induce hæmorrhage. In any case the procedure must be terminated at once if a marked fall of pressure takes place or the general condition of the patient grows rapidly worse. Fortunately brain tumors in children develop very slowly and the child's skull is not very resistant, so that symptomatic operations of this kind are not often called for.

For the rest, the treatment of brain tumor is purely symptomatic. The most important indication is to relieve the headache, and for this purpose all the drugs at our command, particularly antipyrin and also morphine must be employed. In other respects we should apply the same treatment as in any acute process in the brain accompanied by increased pressure.

TUMORS OF THE SPINAL CORD

The pathologic and clinical picture of tumors of the spinal cord in the adult has recently attracted increased attention and has been made the subject of very thorough study. Interest in these neoplasms was aroused by the success achieved by intrepid operators working in conjunction with able neurologists (the foremost among whom were Horsley and Gowers) in accurately determining the seat and then successfully removing a tumor of the spinal cord. Important as is this achievement for adults suffering from this baneful disease, it finds but little application in pediatrics because in the case of children the tumors are rarely extramedullary and sharply circumscribed, being in most cases extensive neoplasms and quite frequently involving the medullary substance of the spinal cord. Since the most common tumors in the spinal cord are tubercles, the same operative difficulties that we have described in connection with brain tubercle are encountered.

Tumors of the cervical canal may be situated (1) in the bone, (2) in the meninges, and (3) in the spinal cord itself.

With regard to the incidence of these tumors, it appears from Schlesinger's analysis that bone neoplasms (carcinoma, sarcoma) are practically never seen in children, with the exception of metastatic sarcomata, which are occasionally encountered. Among meningeal tumors mention should be made of sarcomata, which usually attack the meninges primarily and later spread to the spinal cord and to the brain or appear as secondary tumors in the form of gliosarcoma especially after orbital sarcoma. Lipoma of the spinal meninges is also observed in early childhood and is probably the result of some congenital formation. Within the spinal marrow tubercles are most common in childhood, either single or more frequently in the form of multiple tumors. Primary gliomata, which may become diffuse, are also seen. Gummata of the spinal cord are rare in children and are difficult to distinguish from inflammatory changes in the spinal marrow and from meningitis.

The incidence of spinal tumors during the different periods of life is as follows: in children under ten years of age tubercle is the commonest among intramedullary, as lipoma and sarcoma are the most frequent among extramedullary tumors. In the second decade of life tubercles again preponderate within the spinal marrow, and gliomata are also seen. Outside of the medullary substance, primary or metastatic sarcomatosis is the affection most frequently observed.

The **symptomatology** of tumors of the spinal cord varies with the seat and character of the neoplasm. The widest variations are observed from cases in which the neoplasm either produces no symptoms at all or symptoms that are quite insignificant in comparison with those of a general disease, to the familiar clinical picture of the greatest gravity, presenting signs of a neoplasm of the spinal cord, although the localization is not always easy to determine. The indefinite character of the clinical picture in tumor of the spinal cord is illustrated by a carefully studied case of Heubner's in which there were present paralysis of both legs, with permanent flexion, great pain on passive movement, attacks of tonic spasms in the arms, besides blindness and atrophy of the optic nerve. The autopsy revealed multiple gliomata in the spinal marrow, gliomatous degeneration of the posterior columns, and hydrocephalus, probably caused by an ascending meningitis. In multiple sarcomatosis the course of the disease is sometimes stormy and accompanied by fever.

Aside from these unusual findings the picture of tumor of the spinal cord is about as follows: The first and most important symptom is pain, radiating into one or more extremities according to the seat of the tumor, or in the form of girdle pain. These neuralgias, which are rarer in children than in adults, must be regarded as direct symptoms of irritation of the nerve roots. Hyperæsthesia is sometimes present in the affected part of the body. Motor symptoms of irritation are more rare. They include tonic contractures and possibly direct spasms, limited to one or more extremities. The stage of irritation of the nerve roots may be quite protracted and is always present when the neoplasm begins outside of the medullary substance.

Paralytic phenomena occur both in the sensory and in the motor nerves. Accordingly we observe anæsthesia, limited to one or two extremities or parts of extremities and, on the other hand, loss of power or atrophy in certain definite muscle groups. Depending on whether one half of the spinal cord or the entire cross-section is diseased, there is either a hemiplegia or a complete transverse paralysis. The typical picture of paralysis due to unilateral lesion of the spinal cord is found in so-called *Brown-Séquard paralysis*. In this disease there is motor paralysis on the side of the tumor ,with exaggeration of reflexes and loss of the sense of position, and on the opposite side anæsthesia of all varieties of sensation with the exception of the sense of position. On the side of the motor paralysis a hyperæsthetic zone, which is the expression of nerve root irritation, is not infrequently observed at the height of the spinal disease. In explanation of this paralysis suffice it to say there that the motor disturbances on the same side are caused by disease of the spinal tracts which are uncrossed, whereas the sensory tracts, which are responsible for the sensory disturbances, cross to the other side soon after entering the spinal cord. This Brown-Séquard paralysis is a very characteristic symptom of tumor of the spinal cord, because, with the exception of rare injuries, it does not occur in other diseases of the spinal marrow. Unfortunately, it is not always sharply defined; the sensory paralysis, particularly, is sometimes limited to certain kinds of sensation (pain, temperature).

If the tumor involves the entire spinal cord, the symptoms of Brown-Séquard paralysis become indistinct and the picture of a *transverse lesion* gradually makes its appearance. Corresponding to the most frequent seat of tumors of the spinal cord in the thoracic portion, we find spastic paraplegia with anæsthesia of both legs, disturbance of the bladder and rectum, and decubitus. When the neoplasm involves the cervical or lumbar enlargement, an extremely variable picture may be produced and the symptoms may be exceedingly difficult to interpret, because in these cases, depending on the seat of the tumor, localized muscular atrophies may be added to the sensory and spastic paralysis.

The following points are of importance in determining the level of a spinal tumor.

Tumors situated in the upper cervical portion: At first unilateral paralysis or Brown-Séquard paralysis, later paralysis of all four extremities, pain in the distribution of the cervical plexus, paralysis of the muscles at the back of the neck, with possibly rapid fatal termination from paralysis of the phrenic nerve. *Cervical enlargement:* Flaccid unilateral paralysis of the arm, often spastic paralysis of both legs, paralysis of the muscles of the trunk. If the tumor is not very extensive, there may be only paralysis of the individual muscles of the arm. *Dorsal portion.* Typical picture of Brown-Séquard paralysis, then paraplegia and disturbance of the bladder and rectum. *Lumbar enlargement:* First, unilateral pain in the lumbar plexus radiating into one leg; atrophic paralysis of individual muscles of the thigh and leg; possibly Brown-Séquard's syndrome as regards the two legs; later, complete sensory and motor paralysis of the legs with abolition of the patellar reflex but preservation or exaggeration of the Achilles tendon reflex. *Sacral portion*: Atrophic paralysis of the leg, the foot, the gluteal muscles and the levator ani; loss of sensation in the legs, the inner aspect of the thigh, the foot and the anal region; disturbance of the bladder and rectum; decubitus; loss of Achilles tendon reflex; the knee phenomenon is usually present. As a rule the tumors are so large that they involve both the lumbar and the sacral portions of the cord. *Cauda equina:* Bilateral, rapidly developing paraplegia; intense pain, especially in the sacrum and coccyx; anæsthesia in the region of the rectum.

The **diagnostic difficulties** presented by a tumor of the spinal cord can readily be appreciated from the above description. The important points in the diagnosis are: Onset with pain when the tumor is extramedullary; Brown-Séquard paralysis when the tumor is situated in the medulla; and, finally, the symptoms of a transverse lesion as the entire spinal cord becomes involved. But it must be specially emphasized that small tumors of the spinal cord, and especially tubercles, often fail to give rise to any special symptom sufficiently definite for localization, and frequently escape discovery until they are revealed at the autopsy. The conditions that would be considered in the differential diagnosis, assuming that the possibility of tumor is thought of at all, are: spinal meningitis and spondylitis, the diagnosis of which is rendered possible by the characteristic symptoms which appear later.

The **prognosis** of tumors of the spinal cord is very unfavorable. If the patients do not die of the disease itself, death often results from decubitus and paralysis of the bladder.

Aside from mere symptomatic remedial measures, the **treatment** consists logically in removal of the neoplasm. The difficulties of such a procedure in childhood have already been explained and, as a matter of fact, we know of no case in which a spinal tumor in a child was subjected to operative treatment.

MULTIPLE CEREBRAL AND SPINAL SCLEROSIS

Multiple cerebral and spinal sclerosis consists in the presence of numerous, dense, yellowish white foci in the spinal cord, medulla oblongata and cerebrum. Microscopic examination reveals excessive proliferation of the neuroglia, such as is not attained in any other central disease of the nervous system (Weigert). The nervous tissue itself shows very little change; the axis cylinders of the nerve fibres within the sclerotic patches are for the most part preserved and the ganglion cells are intact. Products of degeneration of nerve substance are found only around the foci. Secondary degeneration of nerve tracts is usually absent.

The above anatomical picture, which has recently been clearly defined by Müller, is to be sharply distinguished from the proliferations of connective tissue which occur after disseminated encephalomyelitis (secondary multiple sclerosis, Schmauss, Ziegler). "Multiple gliosis," as we may call multiple sclerosis on the strength of the anatomical findings, is probably attributable to some congenital condition of the neuroglia. The affection might be classified among the endogenous diseases except that it is not hereditary or familial. In view of the neoplastic, progressive character of the anatomic changes it seems more justifiable to include multiple sclerosis among the neoplasms of the nervous system.

If we accept Müller's theory in regard to genuine multiple sclerosis which we have given above and which is explained in his comprehensive monograph, we must arrive at the very remarkable conclusion that this disease does not occur in childhood. Schupfer in his essay on infantile focal sclerosis demonstrated that the cases which have been described in extraordinary large numbers will not bear criticism. In fact, the very cases in which the triad formulated by Charcot as characteristic of multiple sclerosis, namely, intention tremor, scanning speech and nystagmus, were present turned out, on pathologic examination, to be cases of pseudosclerosis, hereditary syphilis, endogenous degeneration, or cerebral infantile palsy. The very few cases—three in number—which Schupfer accepts as cases of multiple sclerosis presented chiefly disturbances in the motor action of the legs (paraparesis, tremor), disturbances of sensation, defective movements of the eyes, and weakness of the bladder. Hence a diagnosis of multiple sclerosis based on the above-mentioned characteristic triad is not justified in children. Müller even casts a doubt on the authenticity of the cases accepted by Schupfer. He is unable to find in any of these cases the positive signs of exclusive disease of the neurogliar tissue and regards these cases, including the one which Schupfer carefully studied as a paradigm, as disseminated myeloencephalitis. He explains the alleged influence of infectious diseases in the production of multiple sclerosis in children and contends that the nervous effects which follow the infectious diseases are disseminated, chronic inflammations and not primary proliferation of neurogliar tissue or, in other words, multiple sclerosis. Under these circumstances Müller arrives at the conclusion that as yet there is no proof of the occurrence of fully developed infantile focal sclerosis identical with genuine multiple sclerosis and that the cases which hitherto have been regarded as multiple sclerosis on the strength of the anatomical findings merely represent the terminal stages of disseminated encephalomyelitis.

Until this contention, which is based on accurate studies, shall be refuted by arguments based on anatomical findings of an opposite nature, we are not justified in making a diagnosis of multiple sclerosis in the child based on clinical symptoms and are therefore constrained to classify all the cases hitherto regarded as multiple sclerosis under some different head.

SECTION VII.

TRAUMATIC DISEASES OF THE CENTRAL NERVOUS SYSTEM

The etiologic significance of traumatism in the production of nervous diseases in childhood is frequently overestimated by laymen and probably also by physicians. A natural desire to find a cause induces many parents to ascribe nervous diseases in their children to some insignificant accident, such as occurs every day, and they often fall into the error of dating the beginning of the disease from the time of the accident. In the popular mind traumatism plays the same part in the etiology of nervous diseases as catching cold in that of internal diseases. As a matter of fact, the present tendency is to ascribe much less importance to external violence in the production of organic nerve disturbances than was formerly the case, and to give much more prominence to hereditary and family disposition and to bacterial toxins. Nevertheless we have no desire to deny that traumatism is an important causative factor in many diseases of the brain and spinal cord, such for example as tuberculous meningitis, spondylitis and encephalitis; that an injury may lead to suppuration if the wound becomes infected; and, finally, that birth injuries may be responsible for the most severe lesions of the central nervous system, a fact which is only beginning to be properly appreciated (Finkelstein).

1. CONCUSSION OF THE BRAIN (COMMOTIO CEREBRI)

Concussion of the brain is described by Simon as follows: "The condition consists in a contusion, a displacement of the brain as a whole without injury to the brain substance. It results in diminished irritability of all the centres in the cerebral cortex and this loss of irritability may, under certain circumstances, go on to complete functional disability." The symptoms are loss of consciousness, vomiting, slowing of the pulse, retention of urine and, sometimes, transitory palsies and aphasia. Cases with palsy and aphasia justify the assumption of a local contusion of the brain, usually due to contrecoup. In the more frequent milder cases the above-mentioned cerebral symptoms are slight and often disappear before the child is seen by the physician.

Concussion of the brain is not frequent in childhood. In infants particularly it is rarely observed. The reason of this probably is that children rarely fall from a great height and, on the other hand, the childish skull is soft and yielding and therefore offers less resistance to displacement of the brain. On the other hand, on account of the thinness of the cranial bones there is a predisposition to fracture.

The **course** and the **prognosis** in concussion of the brain are on the whole favorable. After a few hours, or rarely days, complete recovery takes place and no permanent disturbances remain as a rule. Cases of severe head injuries are observed in which the brain symptoms gradually increase and the child ultimately dies. In these cases it is natural to suspect a cerebral hæmorrhage and the suspicion is confirmed by the gradually progressive character of the symptoms. However, concussion of the brain may sometimes end fatally without any complications (von Bergmann).

The **treatment** consists in rest, lowering the head, keeping up the activity of the bowels and kidneys (catheter), regular diet and possibly the administration of heart tonics.

2. CONCUSSION OF THE SPINAL CORD

This affection, which has frequently been described in the adult, does not appear to occur in childhood. The condition consists in shock, due to some violent concussion (railroad accident), and produces, in addition to pronounced prostration, temporary and permanent spinal symptoms and possibly paralysis of the bowel. The post-mortem findings in fatal cases were negative as regards the cord, although this has not been accepted without a protest.

3. CEREBRAL HÆMORRHAGE

Intracranial hæmorrhage occurs as the result of a birth injury or a traumatism occurring at some later period.

Among the **causes** of cerebral hæmorrhage after the child's birth, are injuries, whooping-cough, purpura, severe atrophy, and other diseases of the brain, especially sinus thrombosis.

The **symptoms** resemble those of brain embolism and do not differ materially from the symptoms observed in the cerebral hæmorrhages of adults. Convulsions and coma are nearly always present at the beginning. The child either dies with these initial symptoms, or recovery takes place with spastic paralysis, presenting the picture of cerebral infantile palsy. In the main, cerebral hæmorrhages of this kind are rare in children, since the predisposing causes which are present in adults, arteriosclerosis, brain aneurysm and the like, are not operative in childhood.

On the other hand, intrapartum hæmorrhage into the meninges possesses a clinical importance which even yet is not properly appreciated. If we examine post mortem a large number of newborn infants without making any special choice of subjects, we are surprised by the frequency of hæmorrhage within the cranial cavity. There is no doubt that in the majority of cases of otherwise healthy newborn infants the blood is absorbed without producing any clinical symptoms; but in a not inconsiderable minority of the cases the hæmorrhage is of such extent as to render life impossible. Between these two extremes there must be a long series of intermediate degrees, the recognition of which is probably impossible and which no doubt are of great importance in the production of cerebral symptoms that manifest themselves later. As the cause of submeningeal hæmorrhage is by no means clear in every case, recognition of the accident is difficult. While in most cases the occurrence of congestion and laceration of blood vessels within the skull is readily explained by a severe protracted labor with marked displacement of the cranial bones (Kundrat), an easy spontaneous delivery may also lead to submeningeal hæmorrhage (Finkelstein), and this is particularly apt to be the case in premature labors.

In most of the cases the hæmorrhage is situated at the vertex, somewhere in the region of the two paracentral lobules, where heavy deposits of coagulated blood are found. The hæmorrhage may involve one or both convexities of the cerebral hemispheres (Sarah MacNutt), more rarely the base of the skull and the cerebellum. The subjacent portions of the cortex are compressed and infiltrated with blood. So far as I know, no recent cases have as yet been examined by modern histologic methods. In children who have survived a hæmorrhage porencephalus, external hydrocephalus and local meningoencephalitis have been demonstrated as end-products of the lesion.

The **symptoms** of severe intrameningeal hæmorrhage do not always point to the brain. We have instead the picture of severe asphyxia; respiration is either abolished at once or becomes very feeble (atelectasis, Kundrat); the child is cyanotic; and the temperature falls. Death ensues either from gradual failure of respiration or with convulsions. In other cases convulsive seizures, trismus or tetanoid convulsions, spasms, exaggeration of reflexes dominate the picture, and these cases also frequently end fatally. Some infants, however, survive these conditions and actually or apparently recover (Henoch, Finkelstein and others). In a third class of cases the initial symptoms are very slight — no more than a short period of asphyxia, from which the children apparently recover completely. Later, however, convulsions make their appearance in different parts of the body, or without such convulsions the children develop spastic paralysis, which will be described later in connection with cerebral infantile palsy (Little's disease). With what degree of frequency intrapartum hæmorrhage, which produces no symptoms at the time, later leads to epilepsy and idiocy, is difficult to decide. Such an etiologic relationship must be suspected whenever convulsions develop in infants several weeks old without any recognizable cause.

The **treatment** of intrapartum hæmorrhage is the same as that of asphyxia neonatorum.

4. HÆMORRHAGE INTO THE SPINAL CORD

In hæmorrhage into the spinal cord, also, birth injuries are undoubtedly the most important etiologic factors. Schaeffer found extravasations of blood into the vertebral canal in 10 per cent of all his autopsies on newborn infants, and although these figures appear to me somewhat high, I can confirm the frequent occurrence of hæmorrhage of this kind from my own experience. The hæmorrhages are often extraspinal, in which case they are found chiefly on the ventral surface of the lumbar portion of the cord. It is probable that the blood is not always the result of a local extravasation, but consists in part also of blood that has flown down from above after hæmorrhage within the cranium and in the highest portions of the spinal column. Hæmorrhages within the substance of the spinal marrow are extremely frequent in newborn infants. Such hæmorrhages are usually small and of no importance; their favorite seat is at the junction between the posterior and anterior horns. Goldscheider and Flatau's experiments on animals convinced them that fluids injected into the vascular system have a special tendency to escape at this point. Numerous small hæmorrhages into the spinal cord are found especially in premature infants and in anencephalous monsters. Large hæmorrhages into the spinal marrow may conceivably lead to cystic cavities, and the latter may bear some etiologic relationship to a later syringomyelia (Schultze, Zappert and Pfeifer). Hæmorrhages have also been observed in the newborn within the central canal; indeed parts of the canal are sometimes separated by the action of the hæmorrhage.

The above conditions all represent more or less unexpected autopsy findings in children who die soon after birth. General palsies in the newborn can be attributed to hæmorrhage of the spinal cord only in very rare cases (Oppenheim, Raymond).

All other causes play but a minor part in the etiology of spinal hæmorrhage in the newborn. Traumatism is more apt to produce an external injury than an isolated hæmorrhage. In whooping-cough the sudden appearance of a spinal palsy during an attack naturally suggests the probability of hæmorrhage into the spinal cord (Mauthner, Bernhardt and others), but we have no positive autopsy proof that such an accident occurs. Steffen reports a case of hæmorrhage into the spinal cord after purpura.

In cases of this kind the **diagnosis** of spinal hæmorrhage is based on the sudden appearance of symptoms such as have been described in connection with tumors of the spinal cord. Paraplegia, anæsthesia and sphincter paralysis are the most pronounced symptoms in the beginning; they rarely increase during the first few days and are more apt to disappear in a short time. There finally results a clinical picture which corresponds to that of a poliomyelitis, Brown-Séquard palsy, or transverse myelitis. Pain is not common in central spinal hæmorrhages. In this stage localization of the spinal lesion is possible; but unless the history is very precise, the diagnosis of hæmorrhage must always remain doubtful because the ultimate results of inflammation and those of hæmorrhage are clinically identical.

Unless the hæmorrhage proves immediately fatal (hæmorrhage in the upper portion of the cervical cord) the **prognosis** is not altogether unfavorable. But the improvement which at first takes place usually does not go on beyond a certain point, after which one of the above-mentioned permanent conditions develops.

In all cases of hæmorrhage into the spinal cord the **treatment** consists in absolute rest, the application of ice bags, and the exhibition of styptics such as ergotin and gelatin. Catheterization must not be forgotten when there is paralysis of the bladder.

5. FRACTURE AND LUXATION OF THE VERTEBRAL COLUMN

Dislocations and fractures of the vertebral column are exceedingly rare in childhood and differ but little from the same conditions in adults. Occasionally they are observed after instrumental deliveries. I once saw the entire cervical portion separated from the rest of the column at the autopsy on a newborn infant.

The local **symptoms** of a bilateral luxation of the cervical column are: anterior inclination of the head; unusual prominence of one of the spinous processes posteriorly; prominence of the body of one of the vertebræ palpable through the pharynx; fixation of the head; extreme tension of the muscles. In unilateral luxations the head is inclined to the opposite side, the cervical column is convex toward the dislocated side, and the muscles on the side of the luxation are tense. Luxations are seen chiefly in the cervical column, fractures at any point of the backbone and particularly in the lower portion.

The cord itself may escape injury. If the latter is present it is due to the direct pressure of the injured vertebra; to hæmorrhage into the central canal, into the meninges or into the substance of the spinal marrow. Symptoms of hæmatomyelia and other symptoms pointing to the nervous system are not necessarily present. When present, they are usually the same as the symptoms of compression of the spinal cord from caries, or of spinal hæmorrhage, or, if long continued, of spinal tumor. Injuries and luxations of the first two cervical vertebræ are usually rapidly fatal; injuries of the third and fourth are of very bad omen because of the danger to the phrenic nerve. The remaining spinal symptoms can be deduced from the table on page 245. Erection of the penis is one of the commonest symptoms of injuries of the cervical column. The differential diagnosis between luxations and fractures often necessitates an X-ray examination.

The **prognosis** is always extremely grave. If death does not occur in a short time, or, in the case of luxations, if the latter is not successfully reduced, a chronic condition similar to that of myelitis with all its dangers may be expected to develop. For the **treatment** the reader is referred to the sections on surgical diseases.

SECTION VIII.

CEREBRAL INFANTILE PALSY

(*As the terminal condition of various diseases of the brain*)

A number of cerebral affections in childhood, which end neither in complete recovery nor in death, leave behind certain anatomical defects and clinical disabilities. A permanent condition results in which the child is healthy except for certain symptoms which remain of the former disease, so that the condition represents *not a disease but rather the termination of a disease.* This condition is designated *cerebral infantile palsy.* According to the strict acceptation of the term it includes only cerebral affections in which paralysis of the extremities is the most prominent symptom, but there are numerous transitional cases which present merely epilepsy, idiocy or atrophy of the optic nerve, and which are recognized as "cerebral palsy without paralysis" (Freud). It is therefore obvious from what has been said, that the diagnosis of cerebral palsy cannot lay any claim to scientific accuracy, and if we wish to be strictly logical and scientific we ought, instead of speaking of cerebral infantile palsy, endeavor to use such terms as the remains of fœtal diseases of the brain, of cerebral hæmorrhage or of encephalitis. This, however, is as yet impossible because the same etiologic factors do not always give rise to the same clinical disabilities, so that in a given case, it is impossible even with the aid of the history, to arrive at a definite conclusion with regard to the primary disease. We must therefore content ourselves with the well-defined forms of cerebral infantile palsy and are not justified in tearing down the edifice of cerebral infantile palsy, which has been built up by a number of excellent workmen, until a larger amount of building material than we at present have at our disposal has been collected. The diseases which lead to cerebral infantile palsy or, in other words, the etiology of the disease may be classified as follows:

1. *Intra-uterine* (*prenatal*) *causes:* Malformations of the brain and cerebral diseases that are merely concomitants of a general disease (typhoid fever?) or injury to the mother (severe blow on the abdomen). In this class belong cases of porencephalus, microcephalus, atrophy of one hemisphere with unilateral diminution in the size of the skull or depression in certain portions of the cranium, congenital cysts, and the like. Clinically these varieties cannot always be recognized as such, and the diagnosis is often dependent on the history. But one cannot always rely on the history, as most parents fail to recognize congenital motor disturbances in their children until they first attempt to sit up or walk. Sometimes the presence of some other malformation calls attention to the possibility of a congenital disturbance. To what extent congenital porencephalus and microcephalus are to be regarded as due to arrested development or disease of the brain has already been discussed. There are a few cases in the literature which make a relation between intra-uterine traumatism and cerebral infantile palsy probable (Cotard, personal observation). Heredity and alcoholism probably have no etiologic significance; the influence of syphilis has been discussed in another place. The theory that emotional excitement during gestation may have an influence on the child's central nervous system is scarcely tenable.

2. *Birth injuries* (natal, intrapartum causes). These are chiefly intermeningeal hæmorrhages (see section on traumatic diseases of the central nervous system). The term Little's disease is employed by many writers, as he was the first to describe this important cause of numerous cases of cerebral infantile palsy. An intrapartum injury may be assumed a priori if the labor was difficult, or the child was born in asphyxia, or convulsions developed soon after birth. Even when the labor has not been difficult, especially if it was precipitate, intracerebral hæmorrhage is possible. Cases of cerebral palsy occurring after premature deliveries may possibly be explained in this way.

3. *Among extra-uterine causes*, head injuries involving the skull or accompanied by subdural hæmorrhage may lead to cerebral infantile palsy. Circumscribed encephalitis, which may develop spontaneously (polioencephalitis, Strümpell) or in the wake of some infectious disease (measles, scarlet fever, varicella, intestinal catarrah, pneumonia and the like) is a very important cause of cerebral infantile palsy, particularly the unilateral variety. Sinus thrombosis and embolism may produce the same clinical result; the emboli in most cases are probably of infectious or septic nature and are comparatively rare in otherwise uncomplicated cases of endocarditis.

To what extent syphilis may be responsible for the production of cerebral infantile palsy is difficult to say. This etiologic relation was formerly regarded as very rare (Sachs, König), but more recent observations (Rolly, König, Fournier, Erlenmeyer and others) tend to show that permanent cerebral symptoms occur more frequently in hereditary syphilis than was formerly believed. The diagnostic difficulty arises from the impossibility, in the cases of children with hereditary syphilis presenting symptoms of a cerebral palsy, of determining with certainty whether one is really dealing with the remains of a former pathologic process (that is to say, a cerebral infantile palsy, as defined above) or with a temporarily permanent stage of a still active brain lues.

In a large number of cases all we can learn about the beginning of the disease is that the palsy developed within the first two or three years of life after convulsions, which in most cases are said to have been unilateral. Sometimes it is stated that the child had repeated attacks of convulsions before it became permanently paralyzed. In these cases it is not proven that the convulsions produced an alteration in the brain (hæmorrhage) which led to the paralysis; it is quite possible that (prenatal, natal, intrapartum) changes were already present in the brain at an early date and that the motor region became involved secondarily (Freud, Rie).

Finally, there are plenty of cases of cerebral infantile palsy in which, either owing to the indifference of the parents or the very mild character of the initial symptoms, it is impossible to elicit any definite history, so that we can form no conclusion in regard to the date when the palsy began. The existence of this group of cerebral infantile palsy alone precludes a complete etiologic classification of these conditions.

For the study of the **pathologic anatomy** of cerebral infantile palsy we have at our disposal the terminal conditions of those pathologic processes which represent the foundations of the disease that ultimately leads to cerebral infantile palsy. It must be remembered that various diseases are capable of producing the same permanent changes in the brain. The initial lesions, as has been stated, may owe their origin to several different causes, most important among which are meningeal cerebral hæmorrhage, embolism, encephalitis, and intra-uterine diseases or injuries. As a rule, the changes produced by these diseases can no longer be distinguished when the cases of cerebral infantile palsy come to autopsy.

The changes observed at the autopsy of individuals with cerebral infantile palsy, which represent the final results of the morbid process, are:

Porencephalus.—This may be congenital and the expression of a true malformation of the brain or of an intra-uterine disease on the one hand, or may be the result of some process acquired in later life. The defect is found chiefly in the area supplied by the middle cerebral artery and by the artery of the fissure of Sylvius, particularly in the central and temporal convolutions. The time when a given defect in the brain was produced is sometimes very difficult to determine. Either associated with porencephalus or as an independent condition, we also find in cases of cerebral infantile palsy, diminution in the size of the convolutions (microgyria), which in turn is due either to arrested development or to a former inflammatory process. Sachs employed the term *agenesia corticalis,* based on microscopic findings. It is probable that many of these cases are due to secondary changes in the cortex, the result of meningeal hæmorrhages (Oppenheim). Another pathologic condition that belongs under this head is unilateral atrophy of the brain, which is due to sclerosis of one cerebral hemisphere. The histologic changes consist in diffuse sclerosis with proliferation of the neurogliar tissue, thickening of the vessel walls, hypertrophy of the perivascular connective tissue, and diminution of the nervous elements. Diffuse sclerosis may also be bilateral. It has been described as a separate clinical entity which we have already discussed in section V. Localized sclerosis, with bosselated thickening of certain portions of the cerebrum (tuberous sclerosis), has been found at the autopsies of cases of cerebral infantile palsy. The cerebral sclerosis probably represents the end-product of vascular disturbances (occlusion of vessels, inflammation), but gives no clue to the original disease.

Among the anatomical findings in cerebral infantile palsy a place must be reserved for *microcephalus.* As has already been explained, the subdivision of this condition into pseudomicrocephalus (terminal stages of a cerebral disease) and genuine microcephalus (failure to develop beyond a certain stage) is practically impossible. Combinations with abnormal types of cerebral convolutions, particularly the persistence of differentiating fissures (macrogyria), is not infrequent in microcephalus. One also finds cysts, foci of softening—often with hæmorrhagic contents—and connective tissue scars in cerebral infantile palsy, which are to be regarded as localized cerebral processes.

These lesions are all found in the brain cortex, in the white matter of the cerebral hemispheres, and in the region of the basal ganglia.

It would be quite justifiable to include congenital or acquired hydrocephalus as well as microcephalus among the causes of cerebral infantile palsy. As a matter of fact, however, this is not done and it is customary to describe cases of hydrocephalus as a separate clinical group, although they have no better claim to nosologic independence than the cases of cerebral infantile palsy.

Symptomatology.—"The etiology, symptomatology and pathology of cerebral infantile palsy are like three large chains of mountains, and the peaks of one group are connected with the summits of another only by the most difficult passes." Such is the simile with which Peritz graphically describes the difficulties that are encountered when one attempts to bring the symptoms of this disease into harmony with certain definite anatomical changes. We shall therefore practically restrict the present exposition to a mere clinical enumeration of the signs of cerebral infantile palsy, and make only the most cautious attempts to establish a connection between the symptoms, and the localization and nature of the changes existing in the brain.

We distinguish infantile hemiplegia and infantile diplegia, according as the extremities of one or both sides are involved. Hemiplegia is characterized by paresis of one facial nerve and spastic weakness of the arm and leg on one side of the body. The right side is somewhat more frequently attacked than the left. The facial palsy is limited to the middle and inferior branches; it varies greatly in degree and, in older cases particularly, is often barely perceptible. The paralysis is best seen when the child beigns to laugh or cry or speak, and is less distinct during rest and when the facial muscles are in active play. In protracted cases of facial palsy, *spasm of the paralyzed muscles* is sometimes produced, so that the sound side appears more smooth and the picture of a crossed paralysis of the face and extremities is simulated. The arm lesion quite often manifests itself in permanent contractures; the elbow and wrist-joints are flexed and the arm is held close to the body; fixation in extension is more rare. When the paralysis and contractures are of this pronounced type, active movements are practically impossible, and passive movements encounter a violent resistance. In other cases there is only a marked rigidity of the muscles; movements are possible, but they are awkward and ineffective. The paralysis is always most marked in the hand (in contradistinction to spinal palsy). The fingers are folded over the thumbs and it requires considerable effort to open the hand; the finer movements of the hand are performed only with great difficulty. Even when the signs of impaired function are less marked in the arm, the movements of the hand are still distinctly limited. The legs also present a typical spastic hemiplegia, with extension at the hip- and knee-joint and plantar flexion at the ankle-joint. Pes equinus with spasticity is quite often produced. Even when the paralysis is comparatively mild, the disturbance of the gait is quite marked; the child drags the leg and swings it around (circumduction) in bringing the foot forward. When the gait is not especially interfered with paresis,of one leg may reveal itself in the child's inability to stand alone on the affected leg, to hop, or to rise on the toes. When the child sidesteps to the sound side, the paralyzed leg is dragged instead of being lifted clear, because the movement increases the spasm of the adductors (flankgait, Schüller). Partial monoplegia (monoparesis) of one arm or leg does not occur in cerebral infantile palsy, although the condition may be simulated when the paralysis is much more pronounced in one extremity than in others.

Paresis and *spasm* are therefore the characteristic features of cerebral hemiplegia. They may, however, be combined very unequally. Thus, we see cases in which the rigidity of the extremities is pronounced, while the paralysis is quite moderate. Again, it may happen, for example, that the paresis and the increase in the muscle tone are quite marked in the leg, while the arm presents only a slight increase of the reflexes. In short, the involvement of the extremities in cerebral infantile palsy is by no means uniform, and one may see in the same individual in the course of time a diminution of the palsy and, not infrequently, an increase in the contractures.

The *deep reflexes* are always increased. Ankle clonus as well as the Babinsky reflex and Oppenheim's leg reflex are not infrequent. In older cases and in cases in which the hemiplegic phenomena have been less pronounced, a unilateral increase of the deep reflexes is sometimes the only visible sign of cerebral infantile palsy. Diminution or absence of the reflexes is extremely rare and occurs only in exceptional cases that have never been explained. The *skin reflexes* in these conditions are often diminished.

It is not rare for individual *cranial* nerves to be involved in cerebral hemiplegia. The *tongue* may deviate toward the sound side (on account of paralysis of the muscles on the other side of the tongue), indicating involvement of the hypoglossus. Strabismus is quite frequent. Nystagmus, hemianopsia, a tendency to a forced position of the eyes and atrophy of the optic nerve are observed.

Sensory disturbances are rare and, when present, mostly confined to the hands. The disturbance of *stereognosis* which occasionally occurs is perhaps partly referable to the lack of digital dexterity which prevents the child from feeling objects properly.

Motor aphasia as a sequel of a left-sided cortical lesion is conceivable and is in fact sometimes observed. It is to be remembered, however, that we are dealing with cerebral processes engaged in recovery, and accordingly there is a general interference with the function of speech rather than pronounced aphasia; much the same condition, in fact, as we observe in adults after lesions accompanied by aphasia. It is a noteworthy fact that in a child the power of speech may be restored even after complete destruction of the centre of speech in the left side, suggesting the possibility of a vicarious function in the right cerebral hemisphere. In addition to this kind of speech disturbance, due to a focal lesion, children with cerebral infantile palsy quite often exhibit other minor abnormalities of speech which depend in a great measure on feeble-mindedness or idiocy and are sometimes the expression of bulbar disturbances such as will be discussed later.

Atrophy of individual groups of muscles does not occur in cerebral hemiplegia. On the other hand, interference with the growth of the paralyzed side is not infrequent, particularly when the palsies are acquired in early childhood or are congenital. The face may be narrow, producing the impression of a *hemiatrophia faciei;* both the arm and leg may be smaller in all their dimensions, the muscles as well as the bones taking part in the atrophy (this can be demonstrated with the X-rays). The electric irritability of the nerves and muscles in all such cases, however, is normal or increased, and in doubtful cases this symptom can be utilized to distinguish positively between a cerebral and a spinal palsy.

Muscular hypertrophy occurs when the spasticity is very great and particularly in the case of posthemiplegic motor disturbances, which will be discussed separately. It is to be explained as a hypertrophy due to overwork.

Among rare and unimportant symptoms should be mentioned diminution in the size of the breast, the testicle; anomalies in the growth of the hair, the formation of the fingers, eyes, etc., on one side of the body. Epilepsy and idiocy, which are common in cerebral infantile palsy, will be discussed later.

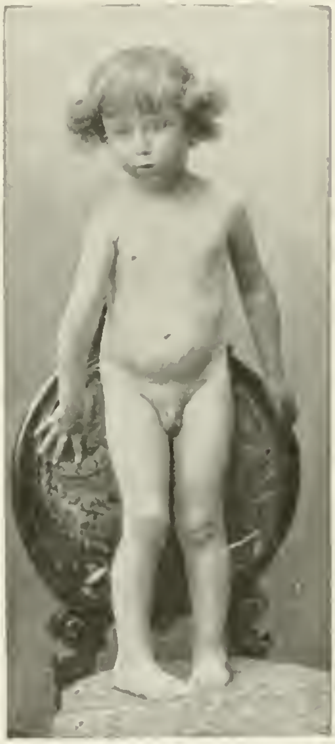
A peculiar phenomenon of infantile hemiplegia is seen in the posthemiplegic motor disturbances, which are of frequent occurrence. The mildest form is the *tremor* which occurs during active movements, particularly at the height of the intended movement (intention tremor); in its severest form the phenomenon consists in *permanent tremor, chorea, athetosis.* These motor disturbances make their appearance either soon after the beginning of the disease or later, following an existing spastic paralysis. The facial muscles sometimes take part in the involuntary movements. In this variety of the disease the paralysis is not marked, but the patients are greatly disturbed by the movements of the arms and fingers which come on with every intentional act. In posthemiplegic chorea the movements are of a jerky, rotating and extending character, and persist practically without interruption except during sleep. In athetosis the characteristic movements consist in spreading, extending and flexing the fingers, and render the child frequently incapable of holding an object in its hands, writing, or doing any kind of manual work. The feet may also take part in these motor disturbances.

Freud describes a peculiar form of motor disturbance under the name of *choreatic paresis.* This differs from the usual form of cerebral infantile palsy by the later occurrence of the disease—after the third year of life, instead of in earlier infancy—by the fact that the motor disturbances manifest themselves at once, and by the absence of spasms, epilepsy or idiocy. In a single case, which was examined post mortem (Landouzy), an old tuberculous nodule was found in the lenticular nucleus.

Cerebral hemiplegia is not always strictly unilateral. In otherwise typical hemiplegic cases rigidity and exaggeration of reflexes, without any distinctly recognizable paralysis, are frequently seen in the leg which is apparently not involved. Such cases form the connecting link between hemiplegia and the second main group of cerebral infantile palsy—*cerebral diplegia.* In the latter the palsy does not exhibit the hemiplegic type; there is either a paraplegia, or all four extremities are attacked. The following varieties are distinguished after Freud:

(*a*) *General rigidity.*—This manifests itself in rigidity of the entire muscular system. The rigidity is noted in earliest infancy and interferes with the necessary manipulations in bathing and dressing the child. Later it is noticed that the child is slow about learning to sit up or walk, and there is a peculiar crossing of the legs when it first attempts to walk (due to great tension of the adductors). The arms are closely pressed to the trunk and flexed at the elbow; the forearms are in pronation, flexed at the wrist; and the fingers are folded, producing a characteristic "devotional or praying" attitude (Freud). The back is rigid, the abdomen hard and retracted, the legs in extreme extension, the feet spastic and in a position of pes equinus. The rigidity is extremely pronounced in the entire body and is much more prominent than the paralysis, which is often very slight. The reflexes, which are difficult to elicit on account of the impossibility of inducing relaxation, are everywhere greatly exaggerated. Sometimes touching the lips, tongue or oral mucous membrane elicits reflex movements of the muscles of the jaw, simulating sucking or masticating movements (Oppenheim's eating reflex). Strabismus and dysarthria are frequent concomitants. The children are easily frightened, especially by sudden loud noises. On the other hand, epilepsy and idiocy are usually absent; although one is very apt, on account of the sluggish movements, slow step and dull expression of countenance, to do the patients the injustice of mistaking them for idiots. This form of cerebral infantile palsy is particularly apt to follow birth injuries and corresponds to the original Little's disease.

Fig. 51.



Cerebral hemiplegia, with posthemiplegic motor disturbances consisting in spastic twitching of the facial distribution on the right side and athetoid movements of the right hand. The illustration shows the hemiplegic position of the right leg. Child four years old.

A special form of this general rigidity is seen in *microcephalic rigidity* in which the skull is diminished in size and there is a high grade of idiocy in addition to the muscular rigidity, which is very marked (see above, microcephalus).

Sometimes the general rigidity is only slight when the child is at rest; but it makes its appearance at once when the child is uncovered, frightened, or hears a loud noise. This condition is called *paroxysmal rigidity.*

(*b*) *Paraplegic rigidity.*—In this form of cerebral diplegia the lower extremities only are rigid and exhibit the same peculiarities as in the cases of general rigidity. The child is very slow in learning to walk and the crossing of the legs at the first attempts is very marked. Paralysis in these cases also is very slight. The arms exhibit at most a slight rigidity and exaggeration of reflexes. The intelligence is normal and convulsions do not occur. On the other hand, strabismus is practically constant. Paraplegic rigidity is observed chiefly in prematurely born children. Opinions are still divided on the question whether this form of spastic paralysis is really due to a cerebral lesion or depends on arrested development of the motor tracts in the spinal cord only (v. Gehuchten). Owing to the frequency of strabismus, however, most authors are inclined to assume a lesion in the cerebral cortex in this form of spastic paralysis also.

(*c* and *d*) When in this form of rigidity the paralytic phenomena are more pronounced, Freud employs the terms *bilateral hemiplegia* and *paraplegic paralysis*, according as both arms and legs or the legs only are involved. The paralysis in these cases may be of unequal degree on the two sides; but in every case the involvement of the hands is very striking, just as in unilateral hemiplegia. The two sides of the face may also be involved, so that the child presents a peculiar, masklike, immobile expression of countenance. Strabismus, convulsions and dementia occur in these forms of cerebral infantile palsy. The cause of these rare disturbances is sought in grave lesions of the brain occurring either before or after birth.

(*e* and *f*) *Bilateral chorea* and *bilateral athetosis* may be present in diplegia as in hemiplegia. In chorea the involuntary movements occur in the muscles of the face, the neck, back and extremities; the patients exhibit persistent, slow rotating and sinuous movements, producing a striking and most alarming clinical picture. Owing to the extreme tension of the muscles of the neck the head is usually retracted. Speech is almost always slow and indistinct. The movements are greatly increased by excitement and when the child knows that it is being observed. The muscular palsies are sometimes quite insignificant and the spasms are not very marked; sometimes there is muscular hypertrophy. As a rule the intelligence is not so much affected as the observer is led to suppose by the slowness of speech and the great interference with movements. Convulsions are absent.

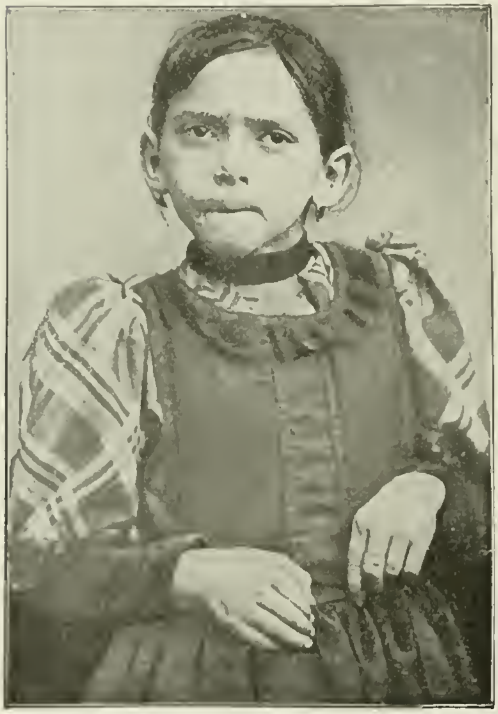
Bilateral athetosis, which is not so conspicuous but none the less disturbing, affects only the muscles of the extremities. The athetosis resembles the above-described unilateral form, but with the difference that both arms and legs are affected. Not infrequently the disturbance is more marked on one side than on the other. Unilateral athetosis may also be associated with bilateral hemiplegia, and in such a case sympathetic movements sometimes occur on the side which is not affected by the athetosis.

Among symptoms referable to the cranial nerves, disturbances of the eye muscles are more frequent in cerebral diplegia than in hemiplegia. Strabismus is the most frequent disturbance: nystagmus is common. Atrophy of the optic nerve may also develop as the result of a neuritis during the original disease. Slowness and difficulty in learning to speak and dysarthria are common in cerebral diplegia; distinct aphasia is rare. Sometimes children with cerebral diplegia appear to be absolutely dumb; but, after a course of systematic speaking exercises, it is often found that inability to speak is due merely to neglect on the part of the child's parents to give it proper training. Apparent deafness may also be due to the same causes. In some cases, however, the inability to hear or speak depends on a high grade of idiocy.

The *bulbar symptoms*, which occur in children with cerebral diplegia and which have been carefully studied by Oppenheim and Peritz and described as infantile pseudobulbar paralysis, merit special attention. The condition is characterized by difficulty in speaking, swallowing and chewing, interference with the finer movements of the lips: partial loss of facial expression; aphonia; and, rarely, by disturbances of the respiration, the heart action, the movements of the eyes and the muscles of the neck. The disturbances manifest themselves only with voluntary movements, the involuntary movements of the affected muscles being preserved. There is no atrophy or fibrillar twitching. The masseter reflex is exaggerated and the eating reflex is often present. In this form of pseudobulbar paralysis the muscles are normally flaccid and the usual muscular tone is preserved. But in addition to the paralytic form there is also a spastic variety in which all the above-mentioned muscle groups persist in a condition of spastic tension and undergo spasmodic distortion whenever a voluntary movement is attempted. An observation of Oppenheim concerning a mother and daughter who both exhibited the same disturbance is frequently quoted. We also know of spurious forms (*formes frustes*) of pseudobulbar paralysis consisting only in slight disturbances of speech, and deglutition. The anatomical basis of this pseudobulbar palsy is not a disease of the medulla oblongata but a cortical lesion involving the centres which innervate the corresponding muscles.

In contradistinction to this form of pseudobulbar paralysis the morbid process (congenital disturbance, inflammation) in genuine bulbar palsy affects the nuclei of the cranial nerves themselves, producing a true picture of ophthalmoplegia or bulbar palsy. Congenital infantile nuclear atrophy is therefore analogous to a congenital defect of the cerebrum and belongs to the group of cerebral infantile palsies. In the same way acute inflammatory disease of the bulb, if it does not terminate fatally or subside completely, may result in recovery with symptoms of permanent loss of function of the bulbar nerves, which properly also belong to the group of cerebral infantile palsies.

Fig. 52.



Cerebral diplegia, with very pronounced spastic pseudobulbar paralysis and athetoid position of both hands. The mother presented the same clinical picture.

The above-described symptoms of cerebral infantile palsy are in many cases associated with two important conditions which are characteristic of this disease, namely, epilepsy and dementia or idiocy.

Epileptic convulsions may be unilateral, and are then always confined to the hemiplegic half of the head; or they may be general. Both forms may alternate in the same individual. The convulsions may occur at long intervals or may be quite frequent and dominate the clinical picture. In most cases they exhibit the typical character of epileptic attacks with the epileptic gait, loss of consciousness, general convulsions, biting of the tongue, etc. The attacks frequently terminate in a definite part of the body. In other cases the convulsions are less pronounced and often consist merely of short attacks of unconsciousness, the conditions described as petit mal or nocturnal epilepsy. There is no constant relation between the intensity of the epileptic attacks and the severity of the disease in the extremities. On the contrary, the cases vary from distinct paralysis or well-marked spasm and slight convulsions to frequent epileptic attacks and almost imperceptible remains of cerebral palsy. Even in the same individual hemiplegia may clear up all but a slight inequality of the two facial nerves of a unilateral exaggeration of the reflexes, leaving only the epileptic condition, which goes on increasing. Indeed, the original cerebral lesion may have been quite insignificant and the symptoms may disappear completely in the course of time while the epilepsy which is probably the result of the irritation caused by the remaining scar persists. Such a condition of affairs is also possible when the primary disease is situated in a "silent" region of the brain where a lesion does not produce clinical manifestations although it is capable of leading to epileptic attacks. On this theory the domain of so-called symptomatic (Jacksonian) epilepsy, in which the convulsions are held to be the result of a circumscribed disease of the brain would be considerably enlarged as compared with that of genuine epilepsy, in which there is no local lesion. It is quite possible, and is strongly suggested by numerous observations, that most forms of epilepsy occurring in childhood belong to the group of symptomatic epilepsies.

Epileptic convulsions are seen chiefly in cerebral hemiplegia, in bilateral hemiplegia, and in microcephalic rigidity. They are less common in universal rigidity, and still more rare in bilateral chorea and athetosis. In paraplegic rigidity and in choreic paralysis they are practically never seen.

The same thing is true of *dementia* and *idiocy*. In this respect also the cases exhibit every grade, from conditions in which the paralytic symptoms predominate, with a mild degree of feeble-mindedness, to those in which the loss of intelligence is very great, while the somatic phenomena are practically absent. In the same way there are all grades of disturbances of the intelligence from feeble-mindedness to absolute idiocy. In most cases the child is merely silly; the more active forms of feeble-mindedness with imperative movements and tic-like habits are rare in cerebral infantile palsy. On the other hand, disturbances of speech and complete inability to speak, as well as masturbation, are quite common in children suffering from cerebral infantile palsy. Defective intelligence occurs chiefly in cases of microcephaly, unilateral and bilateral hemiplegia, and posthemiplegic chorea and athetosis. It is less pronounced in general rigidity and still less in paralytic rigidity. It must not be forgotten that these children, even when the intelligence is not greatly impaired, are in many ways, owing to the motor disturbances from which they suffer, deprived of normal stimuli and of intercourse with other children, and for this reason alone give the impression of being more or less feeble-minded.

Epilepsy and dementia are very frequently present in the same individual, and the same inequality in the intensity of the two phenomena is observed as in the case of palsies. In cases with severe epilepsy a marked degree of idiocy almost always makes its appearance sooner or later.

Finally it should be mentioned that diseases of the eye ground may run their course in a similar manner and that atrophy of the optic nerve may remain as the only symptom of an acute cerebral process —another instance of Freud's "cerebral palsy without paralysis," which was referred to in the beginning of the chapter.

It would be a great advance in the diagnosis of cerebral infantile palsy if it were possible to discover some definite connection between the various clinical pictures of cerebral infantile palsy just described—some of which are very distinctive—and certain anatomical diseases of etiologic conditions. For the present is is impossible, as has already been stated. The following table, in which we have attempted to set forth those relations between the clinical symptoms and anatomical findings which, in the present state of our knowledge, appear to be approximately correct, must not be regarded as more than tentative:

Pathologic Findings	Clinical Symptoms
Microcephalus.	General rigidity, idiocy, convulsions.
Unilateral porencephalus.	Spastic hemiplegia with idiocy, convulsions.
Bilateral porencephalus.	Bilateral hemiplegia with idiocy, convulsions and possibly pseudobulbar paralysis.
Intermeningeal hæmorrhage (birth injury).	1. General rigidity with little or no dementia, usually with convulsions. (Little's symptom-complex). 2. Paraplegic rigidity without dementia or convulsions. 3. Simple hemiplegia with feeble-mindedness, spasms. 4. Bilateral hemiplegia with feeble mindedness, spasms, and possibly pseudobulbar paralysis.
Premature birth (intermeningeal hæmorrhage).	Paraplegic rigidity without dementia, without convulsions.
Head injury (extra-uterine, with hæmorrhage or injury of skull).	Hemiplegia, often with unilateral spasms and feeble-mindedness.
Inflammatory affections in one-half of the cerebrum (also hæmorrhage, softening, etc.).	Hemiplegia with convulsions, feeble-mindedness, posthemiplegic motor disturbances.
Embolism.	Hemiplegic convulsions and feeble-mindedness.
Inflammatory affections in both cerebral hemispheres (also hæmorrhage, softening).	Bilateral hemiplegia with convulsions and feeble-mindedness, possibly pseudobulbar paralysis.
Inflammatory affections in the region of the basal ganglia (also hæmorrhage and softening).	Hemiplegia, possibly hemiplegic chorea and athetosis, choreic paresis.
Inflammatory affections in the medulla oblongata (also hæmorrhage and softening).	Paralysis of the ocular muscles and bulbar symptoms without spasms and without dementia.

The **course** and the **prognosis** of cerebral infantile palsy are such as we should expect from the nature of the disease. Since we are dealing with a reparative process, we do not expect additional focal symptoms to develop but rather look for the further improvement of the palsy. This is what occurs in the great majority of cases and, as a matter of fact, children suffering from severe unilateral or bilateral paralysis are seen to recover. But the improvement is limited; in palsies of the hand particularly recovery is very incomplete. The spasms and the posthemiplegic disturbances exhibit even less tendency to improvement. Although occasionally the rigidity subsides and the muscular tension diminishes, we also see cases in which the contractures go on increasing, and the usefulness of the extremities is severely and permanently impaired. The same is true of choreic and athetoid movements which show no tendency whatever to subside. The arms and hands suffer most in this permanent posthemiplegic condition, as all the finer movements which are necessary for any kind of work appear to be interfered with. On the other hand, children usually learn to walk, though it may be with great difficulty and not without resorting to many artificial aids.

It follows therefore that, in addition to the mild cases in which a relative improvement occurs and the child is ultimately able to work, a large number of individuals with cerebral infantile palsy, particularly with cerebral diplegia, are rendered permanently unable to earn their living. They are often found in hospitals for incurable diseases, in institutions for the feeble-minded and not infrequently in the streets, where their bizarre contortions and strange attitudes excite the pity of the passers-by.

Among the other secondary symptoms of cerebral infantile palsy disturbances of speech and pseudobulbar disturbances also exhibit a tendency to improve. Such patients usually learn to speak in the end, although it may be quite late in life, and their speech may remain permanently defective. Dysphagia also usually undergoes gradual improvement.

The prognosis of *epilepsy* accompanying cerebral infantile palsy is quite as grave as that of so-called genuine epilepsy. The epilepsy manifests itself in a great variety of forms, and its relation to the existing palsies is equally variable, as is also the time of its appearance during the course of cerebral infantile palsy. There are numerous cases in which epileptic convulsions form the opening scene of the clinical picture and remain in the foreground until the end: there are other cases which also begin with convulsions and in which the convulsions later cease; and, finally, there are some in which the convulsions do not appear until years after the paralysis. Unfortunately the epileptic attacks which occur in the course of cerebral infantile palsy have little tendency to subside spontaneously. Much more frequently the attacks increase and often are more prominent in the clinical picture than the palsies. The occurrence of the *status epilepticus* and sudden death is among the possibilities in such cases. Conversely, it is sometimes possible by suitable treatment to convert severe attacks into attacks of shorter duration and lesser severity—into a condition of *petit mal.* The prominent part which epilepsy plays in many cases of cerebral infantile palsy is a warning to great caution in the prognosis of this kind of cases, even when there appears to be some tendency to improvement in the paralysis.

The prospect of *mental development* is not much more favorable than the prognosis generally. Although intellectual improvement takes place in many feeble-minded children and is gratefully accepted by the parents, in most cases of pronounced imbecility the mental impairment is ultimately so great that the child is either unable to go to school at all or goes through with the greatest difficulty, and the question of an occupation becomes a very serious one. Severe grades of idiocy, when associated with microcephalus and unilateral or bilateral hemiplegia, are practically hopeless. It should be reiterated, however, that a great many cases of cerebral infantile palsy present little or no diminution of the intelligence.

We have already mentioned at the beginning of this section that the **diagnosis** of cerebral infantile palsy is purely a clinical diagnosis and leaves the question of the anatomical foundation of the condition open. The history, the absence of progressive character in the paralytic phenomena, and the failure of additional brain symptoms to develop are the determining points for the diagnosis. Hence, slowly developing cases of brain tumor, of family endogenous palsy, may for a time be confused with cerebral infantile palsy, and brain syphilis may temporarily or permanently simulate the picture of a cerebral infantile palsy. Spinal and peripheral palsies are distinguished by the atrophy, which is limited to definite groups of muscles, the presence of the reactions of degeneration, and absence of the tendon reflexes. Posthemiplegic chorea and athetosis could hardly be mistaken for genuine chorea or tic if the case is under observation for any length of time. On the other hand, the physician should be on the lookout for the remains of cerebral infantile palsy in all apparently uncomplicated cases of epilepsy and idiocy.

We know of no **treatment** against the cause of cerebral infantile palsy. Cases that respond to iodine and mercury are suspicious; they are probably brain syphilis and not cerebral infantile palsy. The symptomatic treatment consists in electricity, massage and gymnastics.

The *electric* treatment must be adapted to the peculiarities of the individual case. If the palsies are the most prominent symptoms, the faradic current or the active cathode is employed as in a case of spinal palsy (see poliomyelitis). The electric treatment must be strictly limited to the paralyzed group of muscles, to the exclusion of the antagonistic muscles which are usually hypertonic. If the spasms or posthemiplegic motor disturbances are more prominent, sedative treatment with the anode is indicated; the anode being either lightly passed over the muscles (labile application) or applied to the end of the extremity (stabile application). The cathode rests on the back or the upper portion of the extremity.

Massage, particularly when combined with passive movements and gymnastic exercises, is much more useful than electricity and is specially adapted to cases of cerebral infantile palsy. It acts very well in overcoming beginning contractures, especially when combined with the use of orthopedic apparatus. Warm baths in most cases are followed by subjective and objective improvement of the motility and, combined with artificial movements, are to be recommended. Aeratothermal, saline and carbonic acid baths and sea bathing in the Baltic or Adriatic Seas may also be recommended for children suffering from this disease.

In general all cases of cerebral infantile palsy in which the prognosis is not clouded by the existence of a high grade of idiocy or of epilepsy require medicinal treatment for a long time and constant alternation of the various procedures employed.

Quite recently the transplantation of tendon has been tried in this disease (see poliomyelitis). By dividing individual, greatly contracted muscles and ingeniously changing the point of insertion, a modification of the muscular mechanism may be brought about.

The medicinal treatment of epilepsy and the educational management of imbecility are discussed under the respective diseases.

SECTION IX.

DISEASES OF THE PERIPHERAL NERVOUS SYSTEM

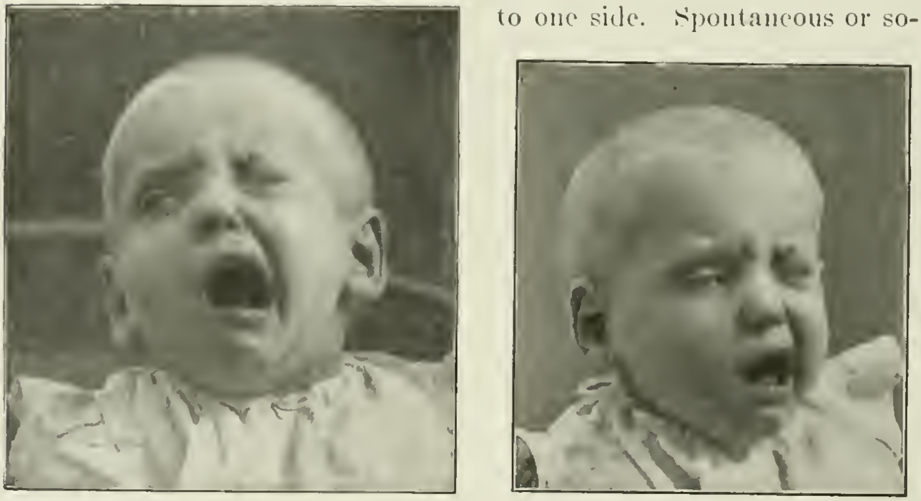
The peripheral diseases of the nervous system and more particularly the palsies, which chiefly occupy our attention, might also be classified according to the etiology. But we do not think such a classification is indicated, partly because for practical reasons a classification according to the seat of the peripheral nerve lesion is more useful, and partly because an etiologic classification would lead to much repetition, as certain palsies which are clinically identical, as for example, facial palsy, may have different causes. Nor shall we attempt in this section to give an exhaustive description of the peripheral nerve palsies, but shall be content to select only those which are important in childhood.

I. PALSIES

1. *Facial Palsies*

A facial palsy is termed *peripheral* when the lesion affects the nerve at some point between the pons and the distribution of the nerve on the face; hence disease of the base of the brain may also produce a peripheral facial palsy. There are cases of partial facial palsy in which only certain muscles of the face are involved. Facial palsy is not rare in childhood since in addition to the "rheumatic form" which occurs spontaneously we also encounter congenital and otogenic lesions of the facial nerve. "Congenital" facial palsy is generally caused by a birth injury, contusion or laceration of the trunk of the nerve in the face by the pressure of the forceps or some obstacle during the passage of the head through the pelvis. For genuine congenital facial palsy, which may be bilateral (diplegia facialis), see the chapter on "infantile nuclear atrophy." A particularly frequent form of facial palsy is confined to the inferior branch of the nerve and leads to a permanent deformity in which the mouth is drawn over to one side. Spontaneous or so-called rheumatic facial palsy is less frequent in the child than in the adult. The possibility of complete facial palsy, accompanied by fever and "exhibiting the signs of a peripheral or pontine paralysis," is mentioned by Oppenheim. The most frequent cause of facial palsy in a child is found in diseases of the ear, not so much simple purulent middle ear catarrh as a destructive bone process (caries of the mastoid bone), which quite often rests on a tuberculous foundation. Diseases of the base of the brain (tumor, meningitis) rarely produce an uncomplicated facial palsy, other basal palsies and cerebral symptoms being usually associated. Finally we may occasionally see a facial palsy, usually partial, after the removal of glands, tumors and the like in the face and at the angle of the jaws.

Figs. 53 *a* and *b*.

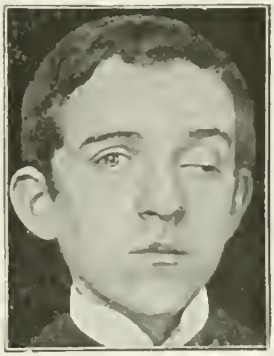


a *b*

Peripheral facial palsy. Child two and a half years old. The frontal branch is involved and the eye remains open during strong contraction of the facial muscles, all of which is well shown in the illustration.

A pronounced peripheral facial palsy differs from the central form by the fact that it involves not only the two inferior, but also the frontal branch of the nerve. The diseased half of the face is flaccid, the nasolabial fold obliterated, the corner of the mouth drawn toward the sound side; the forehead is smooth on the paralyzed side and the eye cannot be completely closed (lagophthalmus); when the child closes its eyes the globe is rotated under the upper lid. The inequality on the two sides of the face becomes very evident whenever the facial muscles are brought into play. Electric irritability is diminished or reaction of degeneration in the form of sluggish, and sometimes inverse contractions are present, or the muscles fail to respond altogether to currents of bearable severity. The prognosis in regard to recovery is determined by the nature of the electric reactions during the first two weeks after the beginning of the palsy. Diminution in the secretion of saliva and the lachrymal secretion and loss of the sense of taste are not infrequently observed. The insufficient closure of the lid during sleep often leads to irritation of the conjunctiva. Pain and paræsthesia are rare in simple facial palsy, but we sometimes observe œdema and herpetic eruptions on the paralyzed side.

Fig. 54.



Periodic oculomotor paralysis during an attack.

Facial palsy due to some birth anomaly almost always runs a favorable course. With few exceptions, the cases recover in a week or two. The rheumatic form also frequently ends in recovery. On the other hand, facial palsy due to aural diseases runs a less favorable course because the primary cause does not subside so soon. Variations and remissions after apparent recovery are observed. But ultimately these cases also not infrequently end in recovery. Sometimes contractures develop in the muscles of the diseased side and make it difficult to decide which side is paralyzed. Since in cases of basal palsy and those due to diseases of the ear, the facial palsy is only a symptom of a more extensive disease the general health of such patients is impaired and they may succumb to the primary trouble.

It is important both from the diagnostic and prognostic viewpoint to determine the seat of the lesion in the peripheral portion of the facial nerve. If the lesion is at the base of the brain, there will be involvement of other nerves (auditory) and symptoms of cerebral disease. If the nerve is diseased in the region of the geniculate ganglion, there may (possibly) be a disturbance of lachrymation and (less certainly) paralysis of the soft palate. A disturbance within the ear produces characteristic symptoms because the corda tympani is given off in that situation. There is impairment of the sense of taste in the anterior portion of the tongue and the secretion of saliva is decreased. In (rheumatic or traumatic) lesions situated at the stylomastoid there is muscular palsy without secretory or gustatory disturbances. The diagnosis between central and peripheral facial palsy rests on the fact that in the former the superior branch usually escapes, on the preservation of electric irritability, and on paralysis of the extremities, which is usually present also.

The **treatment** of facial palsy is chiefly concerned with the cause of the condition. Unfortunately even the complete cure of aural disease by a radical operation is not always followed by recovery from the facial palsy. In fact, the latter not infrequently first makes its appearance after an operation on the ear. In other forms of facial palsy potassium iodide or one of the salicylates (aspirin), depending on the cause, may be administered with doubtful chances of success, and electric treatment resorted to early. Local bleeding behind the ear and hot applications are also emyloyed. If there is lagophthalmus, the eye should be covered with moist dressings during sleep in order to prevent excessive drying of the conjunctiva.

2. *Palsies of Other Cranial Nerves*

Other cranial nerve palsies, as paralysis of the spinal accessory, trifacial and hypoglossus, occur only in association with certain affections of the brain and medulla, so that it seems superfluous to discuss them at this place.

Isolated palsies of the *ocular muscles* may occur and have already been described as congenital, as muscular disturbances, and as the results of hereditary syphilis.

Periodic oculomotor palsy is a term used to describe a disease occurring in children and characterized by paroxysmal, total and partial paralysis of one oculomotor nerve accompanied by migraine. During the intervals between the attacks the muscular function of the affected eye is either normal or but slightly impaired. The individual attacks last only a few days, or, in rare cases, weeks or months. With the exception of neuropathic symptoms that are occasionally present, the patients are otherwise free from nervous disease.

The nature of periodic oculomotor palsy is still in doubt, and opinions are divided in regard to its exact nosologic definition. In the limited number of cases in which autopsy could be performed, neoplasms or inflammations of the oculomotor were found. It is possible, however, that the condition begins as a vasomotor disturbance, which later forms the starting point of more serious lesions. To what extent periodic oculomotor palsy may be classified under the head of migraine is still undecided (Möbius). The prognosis as to recovery is doubtful, and not altogether favorable with regard to the general health on account of the possibility of a neoplasm developing. The *treatment* is the same as that of migraine.

3. *Serratus Magnus Paralysis*

Paralysis of the serratus magnus or, in other words, an isolated paralysis of the long thoracic (posterior thoracic ? Morris) nerve has been observed after injuries, diphtheria, influenza, etc. Possibly it may also be congenital. The most striking symptom is the wing-like projection of the shoulder blade when the arm is raised and brought forward. During rest the shoulder blade is higher than its fellow and the distance between the angle and the vertebral column is diminished. This approximation of the scapula to the median line becomes more distinct when the arm is raised to the horizontal position. The patients are unable to carry the arm beyond that plane.

4. *Palsies in the Distribution of the Cervical Brachial Plexus*

Palsies in the distribution of the cervical plexus are of great importance in childhood; they result from birth injuries (see diseases of the newborn).

The sensory and motor disturbances which result from pressure of cervical ribs on the nerves of the arm usually manifest themselves after the age of childhood.

A disease which is peculiar to childhood, although it is still a matter of dispute whether it should be included among the peripheral nervous diseases, is known as *paralysie douloureuse Chassaignac* or painful paralysis of the arms in small children. It is observed in children between the ages of one and four years and is caused by a sudden pull on the arm as, for instance, in saving the child from a fall, or by some other forced movement of the arm. It is doubtful whether this form of palsy ever occurs without a preceding traumatism; in such cases there must always remain a suspicion that the nurse has probably concealed the cause. The injury is followed immediately by evidences of acute pain and the arm drops to the side as though paralyzed. The physician, who is usually sent for at once by the frightened attendants, finds on examination complete loss of movement in the arm, which is in pronation, but no other disturbances. Passive movements elicit intense pain and resistance on the part of the child, but can be carried out without difficulty in all the joints of the body. No injury is found in the bones or joints, the muscles react normally to the electric current and the deep reflexes are not altered. Examination of the injured arm is rendered difficult on account of the great pain, which is most marked during attempts at supination and pronation; but there are no regular, recurrent painful spots or pressure points. It is difficult to decide whether there is really abnormal sensitiveness to pain in the skin or whether the child cries out when one attempts to touch it because it is afraid of being hurt. The whole condition, which at first is so alarming, rapidly subsides without leaving any permanent damage. In from one to two days motion returns; in from four to five days, sometimes earlier, the normal conditions are entirely restored.

This peculiar disturbance, which was first described by Kennedy in 1850 and six years later in more detail by Chassaignac, has since been made the subject of numerous investigations. While the various authors agree in their description of the disease itself, they offer a variety of explanations; the majority are of the opinion that the disturbance is due to a psychic or inhibition palsy (Vierordt) and that the children avoid moving the arm because they remember the initial pain long after the part has ceased to be painful (Brunon, Laborde, Vierordt). Other authorities, on the other hand, adopt Chassaignac's original theory of a local lesion to the plexus. Bezy and his pupils, Charpy and Abelous, attempted to prove by experimentation, on the one hand, that rapid elevation of the arm causes excessive stretching of the brachial plexus or of certain portions of the plexus, and, on the other hand, that stretching the nerves in the same way in animals produces paralytic phenomena similar to those which are observed in the *paralysie douloureuse* of children. A number of investigators seek the cause in some injury to the joint (partial luxation). Among these are Goyrand, Guersant, Moreau, and Ollier. The last-mentioned author particularly assumed what he called *entorse juxtaépiphysaire, i.e.*, distortion of the epiphysis with laceration of the periosteum. All these theories still have their adherents to the present day, and in spite of Bezy's experiments we find in the latest papers on this subject the theory of a plexus lesion (Lövegren) in sharp contrast to the theory of an inhibition palsy occurring in a neuropathic child (Galatti).

The conditions must be differentiated from actual injuries to the arm (especially fracture of the clavicle) and poliomyelitis. In the former the skiagraphic findings are positive, while poliomyelitis is characterized by the absence of pain; in both conditions the clinical course is characteristic. No *treatment* is required, but I am in the habit of bandaging the arm and believe that by doing so I obtain more rapid healing.

A similar condition has also been observed in the leg (Chassaignac and Brunon).

5. *Palsies of the Nerves of the Arm*

Paralysis of the extensor muscles, with typical wrist drop, flexion of the fingers on the hand, and absence of movements of extension and supination, are sometimes observed in children in cases of peripheral palsy of the radial nerve, which may be congenital or acquired.

The congenital form may be due either to the pressure of an amniotic band, in which case pressure marks may be seen on the arm at the point where the radial nerve passes around the member (Spieler) or it occurs without any recognizable cause, in which case it may be the result of intra-uterine or intrapartum pressure on the radial nerve (personal observation). The prognosis in the former case is unfavorable; in the second the paralysis disappears in a few weeks. These congenital palsies of the radial nerve are usually unilateral.

In later childhood radial palsies may result from lead poisoning or traumatism. Neuritis due to lead poisoning will be discussed later. Pressure (pressure palsy during sleep), unusual muscular efforts, overexertion and other causes of radial palsy which are important in adults are less prominent in children, although they may become operative in fractures of the humerus and during anæsthesia. The occurrence of traumatic radial palsy in children, however, is very rare.

Peripheral palsy in the distribution of the median and ulnar nerves are exceptional conditions, which are extremely rare in childhood and do not differ from similar conditions in the adult.

6. *Palsies in the Lower Extremities*

Peripheral palsies in the lower extremities are much more rare in children than palsies in the upper extremities.

In rare cases a leg may be involved in a birth injury, resulting in crural palsy affecting the muscles of the thigh.

Isolated affections of the peroneal nerve resulting from traumatism may also be mentioned. While it is a well-established fact that the peroneal muscles as well as the tibialis anticus may be affected alone in poliomyelitis and in all toxic neuritides of childhood, no satisfactory explanation has as yet been offered. There is footdrop and, in cases with contracture of the muscles of the calf, the member is spastic and assumes the position of pes equinus, cannot be elevated and in walking the toes drag along the ground.

Unilateral and bilateral club foot is a very common congenital anomaly in the lower extremities. There is neither palsy nor atrophy of the affected muscles, and no pathologic changes are found either in the peripheral or in the spinal nervous system. Whether club foot is caused by a primary muscular action (contraction of the tibialis anticus) or by the position of the child in utero is undecided. In favor of the latter we have the fact that in newborn infants depressions are sometimes seen on the legs or abdomen and that, by fitting the club feet into these depressions, a faulty intra-uterine position may be produced.

Whether *meralgia parœsthetica*, consisting in diminished sensibility in the distribution of the external femoral cutaneous nerve with paraæsthesiæ, occurs in children I do not know. Since the affection occurs with flat foot, it is reasonable to suppose that it might develop in later childhood.

Whether the condition described by Oppenheim as congenital myotonia is nervous or muscular in origin is still doubtful. There is congenital flaccidity and immobility of the muscles of the leg, and more rarely, of other regions of the body, associated with diminution of electric irritability and absent or diminished tendon reflexes. The disease is congenital and, especially when treated by electricity, tends toward rapid improvement. Oppenheim attributes this form of palsy to delayed development of the muscles and possibly to a functional weakness of the motor cells in the anterior horns.

The pseudoparaplegia of rachitic children (Comby, Vierordt) has never been fully explained. The condition is observed in older rachitic children and manifests itself in sudden inability to walk. The legs are flaccid and there is muscular atony; the tendon reflexes and electric irritability are usually preserved. The power of walking is restored after a few weeks even without electric treatment. Whether the pseudoparaplegia is due to a reflex palsy (Vierordt) from the painful condition of the bones or to increase of the normal muscular flaccidity observed in rachitic subjects or, finally, represents a nervous phenomenon, is as yet undecided.

II. DISEASES OF THE SYMPATHETIC NERVE

Paralysis of the sympathetic nerve (contraction of the pupil, and of the palpebral fissure, sometimes retraction of the eye, redness and anidrosis of the affected half of the face), and irritation of the sympathetic (dilatation of the pupil and of the palpebral fissure, sometimes exophthalmos, and hyperidrosis of the same half of the face) are but rarely observed in children either alone or in association with other nervous diseases. According to Oppenheim hereditary palsy and congenital weakness of the sympathetic nerve are possible. The condition may also be produced by the pressure of a tumor in the neck (thyroid gland, lymph-gland, etc.) or by operative traumatisms. The symptoms are variable and the clinical picture is the same in the child as in the adult.

Phenomena referable to lesions of the sympathetic are observed as secondary symptoms in diseases of the spinal cord and of the brachial plexus when the first dorsal root (and the eighth cervical root) which contain oculopupillary fibres passing from the spinal cord to the sympathetic, are involved. Such a lesion produces so-called Klumpke's palsy, characterized by paralysis of the small muscles of the hand and flexors of the forearm, loss of sensation in the distribution of the ulnar nerve on the one hand, and sometimes symptoms of paralysis of the sympathetic on the other hand. In palsies of the extremities due to injury of the inferior trunk of the brachial plexus, particularly in birth palsies, these oculopupillary symptoms are always present. Peters observed similar sympathetic symptoms in syphilitic pseudoparalysis, but his statements have so far not been confirmed.

Anomalies in the secretion of sweat are usually included among diseases of the sympathetic, although the pathologic connection is by no means clear.

It is well known that some persons perspire much more freely when under the influence of heat, excitement and exertion than others, the difference being due to individual as well as familial peculiarities. The tendency to hyperidrosis may be compared to the tendency to blush, which is also very frequently hereditary. Under pathologic conditions disturbances of the sweat secretion may occur both as the symptoms of other diseases and as independent anomaly. Usually there is hyperidrosis—which is much more common than anidrosis or the absence of sweating—in circumscribed portions of the body surfaces.

General hyperidrosis occurs in certain neuroses (general neurasthenia, hysteria) in Basedow's disease, paralysis, in the course of epilepsy, or as an "equivalent" of an epileptic attack. Localized hyperidrosis is seen in cerebral affections (hemiplegia), disease of the spinal cord (poliomyelitis, syringomyelitis, tabes), in injuries of the peripheral nervous system (nerve injuries, polyneuritis), and in diseases of the sympathetic.

Anomalies of the sweat secretion are regarded as idiopathic when no other disease of the nervous system is present and when the irritation which causes the sweating, or the localization of the secretion of sweat, or its intensity, or any or all of these factors are abnormal.

Localized eruptions of sweat in spots or involving one half of the face are frequently observed in children during the act of chewing acid, highly seasoned, or even normal food. The mere sight of such food may call forth the secretion. The corresponding part of the face becomes red, and perspiration appears in large drops on the surface of the skin. It appears that sometimes no more than a violent hyperæmia is produced. The latter phenomena, like most idiopathic anomalies of the sweat secretion, may be observed in several members of the same family. Both the exciting stimulus and the localization of the secretion are abnormal.

Another anomaly of the sweat secretion consists in so-called paradoxical sweating. The subjects of this anomaly perspire under conditions which ordinarily inhibit perspiration, such as cold, while heat arrests the secretion; hence in these cases the stimulus is abnormal. The localization of the sweat secretion may also be abnormal. In these cases of paradoxical sweating, certain portions of the body only begin to sweat under the influence of cold, and in some cases perspiration appears only in parts of the body which generally are not prone to perspire, while the areas where sweating is normally most profuse, such as the palms of the hand, remain dry.

In another group of hyperidrosis we have so-called acrohyperidrosis, a condition in which, after a very slight psychic impression the tip of the nose, the forehead, and the hands or feet may break out in perspiration; in this condition both the stimulus and the degree of reaction must be regarded as abnormal.

Finally there is an anomaly which consists in unilateral hyperidrosis occurring under the influence of heat, the warmth of the bed, or emotion. While the stimulus that produces perspiration is normal in this condition, the localization of the sweat secretion is abnormal.

With few exceptions these anomalies of the sweat secretion are permanent conditions; they are often very troublesome but do not materially affect the general health, although many of them may seriously interfere with the subject's occupation and his social life. These forms of hyperidrosis may be regarded as sudoral reflex neuroses, and it may be assumed that the reflex centres are situated in the spinal cord. The presence of sweat centres in the spinal cord has been definitely established.

The pathology of sweat secretion both in the child and in the adult is in need of further study.

III. NEURALGIAS

All the authors who have made a study of neuralgias agree that they are exceedingly rare in childhood. In a series of 150 cases observed by Remak, only one occurred during the first, and six during the second decade of life. The cases analyzed were undoubtedly cases of typical chronic ("stationary") neuralgias, with paroxysms of raging pain, which children fortunately escape. On the other hand, in my own experience the occurrence of attacks of mild, persistent pain lasting a few weeks is not infrequent among children. The nerves involved are chiefly the trifacial and occipital. The commonest causes of these neuralgias are influenza and coryza. Undoubtedly a nervous disposition is an important etiologic factor. The cases of neuralgia which have come under my observation were almost all in children with distinct neuropathic tendencies, in whom there appeared to be a disproportion in the intensity of the attacks of pain and their complaints. Nevertheless, I am not willing to believe in a simple pseudoneuralgia or psychalgia (Oppenheim) in these cases, or in other words, that the pain is purely psychical; the sudden onset of the disease, which usually lasts but a short time, distinctly points to a local lesion. It is possible that supraorbital neuralgia and the rarer infraorbital form are caused by inflammations in the accessory cavities of the nose. The distribution of the pain in facial and occipital neuralgia in the child, as in the adult, corresponds exactly to the nerve paths, and pain on pressure at the point of exit of the nerves is also observed. Other local symptoms due to vasomotor disturbances or to pain I have never observed in the neuralgias of children. In some cases, even when there is no suspicion of malaria, the neuralgia comes on at a definite time of the day and lasts from a half hour to an hour. Occasionally we observe periodical remissions and recurrences of these neuralgic stages; in these cases it is impossible to deny that the pain may rest on a purely psychic foundation.

The **prognosis** in facial and occipital neuralgia is therefore very favorable in childhood. Cases with persistently recurring, uncontrollable attacks of pain, which belong to the most painful diseases that man is subject to, are fortunately unknown among children. Unless the neuralgic attacks depend on some other progressive disease, they disappear after a few days or weeks or rarely several months.

In the **treatment** of facial neuralgia in children we do not, therefore, need to resort to heroic operative procedures; laxatives, internal medication and electric treatment are usually sufficient. Of the various nerve remedies quinine, antipyrin, aspirin and phenacetin, either alone or in combination, offer good results. Electric treatment consists in the use of the stabile anode and the faradic brush.

Neuralgias in other nerve territories are extremely rare in childhood. Thus sciatica, the most frequent form of isolated inflammation of nerves in adults, is unknown in the child. With regard to various painful conditions in the lower extremity which have recently been carefully studied in adults, such as achillodynia (pain in the tendo Achillis), metatarsalgia (pain in the region of the fourth metatarsophalangeal articulation) and coccycodynia (pain in the region of the coccyx), but little attention has been paid to these things in children. In my experience they occasionally occur in older children.

The occurrence of typical Head's zones in children after the sixth year of age was proved by Bartenstein, who investigated a large series of cases. Picking up a fold of skin or stroking the skin with the head of a pin reveals the presence of painful areas on the trunk. They occur in connection with visceral diseases, and in their localization exhibit a certain constant relation to the various internal organs. The existence of these painful areas is explained by assuming that the irritation affects those spinal roots which give origin to the nerves of the corresponding organ. This theory explains many hitherto inexplicable pains occurring in the course of internal diseases, such as intercostal pain in pneumonia, and at the same time furnishes a theoretical foundation for the various procedures of counterirritation which have long been in use.

The peculiar relationship existing between swelling of the erectile tissue of the nose and uterine pain (Fliess, Schiff) is probably explainable in the same way. In cases of severe dysmenorrhea occurring in girls near the age of puberty, cocainization of the nose, which often proves successful in such cases, may be considered.

IV. POLYNEURITIS

Polyneuritis or multiple neuritis is seen in children after acute infectious diseases and poisoning, or as an idiopathic disease. From the cases reported it appears that diphtheria, scarlet fever (Seifert, Baselli, Remak), mumps (Joffroy), whooping-cough (Möbius), influenza, pneumonia and typhoid fever predispose to multiple neuritis. The most important poisons are alcohol, lead and arsenic. As an independent disease multiple neuritis has been observed not only sporadically but also in epidemic form as well as in association with poliomyelitis and polieneephalitis.

We will first consider the symptoms of multiple neuritis such as they appear especially in the idiopathic form, after which the somewhat aberrant course observed in postdiphtheritic, alcoholic, saturnine and arsenical neuritis will be discussed.

Multiple neuritis (except after diphtheria, see below) is a very rare disease in childhood, particularly in comparison with acute inflammation of the brain and spinal cord, which are so frequent at that period of life. The disease usually begins with weakness and pain in the extremities; the legs are generally attacked first. The child is easily fatigued, there is motor weakness, emaciation, especially in the muscles of the leg, and the deep reflexes are abolished. As the peroneal muscles are chiefly attacked by the disease in children, the gait, aside from the general weakness of the legs, becomes characteristic and is known as stepping gait. Electric irritability in the diseased muscles is usually diminished; sometimes the reactions of degeneration are present. Pain may either occur spontaneously in the legs, or may be elicited by pressure on the nerve trunks and muscles; widespread or localized hyperæsthesia, more rarely anæsthesia, is frequently present. The skin reflexes are also, as a rule, diminished. In most cases the arms are attacked at a later period, when the paralysis of the legs is already quite marked. Again, the forearm and the hand are the parts chiefly involved; the muscles supplied by the radial nerve being frequently involved more than any others (individual muscles in the distribution of the diseased nerve may remain intact). Atrophy, electric irritability, the state of the reflexes, and the sensory disturbances are exactly the same in the arms as in the legs; but the motor and sensory paralysis in the arms is often much less marked than in the lower extremities. The paralysis of the arms is sometimes accompanied by tremor. The palsy is always bilateral and usually uniform in intensity. Extension of the motor weakness to the muscles of the trunk is rare, weakness of the sphincters still more rare; sluggishness of the bowels is somewhat more frequent. The cranial nerves (ocular muscles), the pneumogastric and the phrenic are attacked only in exceptional cases in children. Trophic changes such as abnormal sweat secretion, œdema and changes in the skin sometimes occur. The occurrence of a polyneuritic psychosis (Korsakow) has, so far as I know, never been observed in children.

Multiple neuritis does not always exhibit the distribution described above; it may be limited to smaller territories (legs). Thus I have seen a bilateral paresis of the peroneal muscles develop spontaneously with pain, which could be interpreted only as a neuritis.

Multiple neuritis may come on suddenly like a febrile disease and run an acute febrile course. Insidious onset, however, is more frequent; the disease reaching its acme in from two to four weeks, and persisting at that point for several months. In favorable cases multiple neuritis in children lasts from several weeks to six months. Localized muscular atrophy and paralysis may persist after the disease has run its course. Death is very exceptional, although the primary disease to which the polyneuritis is due may terminate fatally.

The **pathology** of multiple neuritis, in so far as the findings in adults can be applied to children, shows inflammatory degenerative disease of the peripheral nerves. It is not always easy even in the histologic picture to distinguish polyneuritis from nerve degeneration. Spinal changes in the form of poliomyelitic foci and degeneration of ganglion cells and fibres, which may also be present, must be ascribed to some toxic affection of the spinal cord. The changes in the spinal cord must be regarded as coördinate with the peripheral changes and not as the primary condition.

(*a*) *Postdiphtheritic Paralysis*

Characteristic paralytic symptoms occur after diphtheria both in children and in adults. Whether adults are more disposed to postdiphtheritic palsy than children, as has been supposed on the strength of certain not quite uniform statistics, must remain undecided for the present. At all events postdiphtheritic palsy is very frequent in children. Heubner observed paralysis in five per cent., and Goodall in about eleven per cent. of their cases of diphtheria, and these figures are probably too low; for in the public clinics of large cities children do not consult the same physician for nervous symptoms as for diphtheria. The question can be cleared up only by statistics carefully compiled from private practice. It has recently been asserted in many quarters that the number and severity of the cases of paralysis have increased since the introduction of antitoxin. Although it cannot be said that the statement has been definitely proved, it is nevertheless quite plausible when it is remembered that the antitoxin treatment often effects a cure at a stage of the disease when the intoxication of the tissues is so severe that death would have resulted without it.

We distinguish an early and a late form of diphtheritic palsy. In the early form the paralysis—the palate is always affected first—comes on immediately after the angina, so that it is often at first difficult to decide whether the dysphagia is due to the acute pharyngeal disease of whether it is already the effect of paralysis of the palate. More frequently the paralysis is delayed until the second or third week, when the diphtheria itself has run its course and the children are already considered well. I have also observed both forms of palsy separated by a short interval of freedom.

The most important symptoms are: (1) paralysis of the palate, which manifests itself in nasal speech, insufficient closure of the larynx during deglutition, and the regurgitation of fluid through the nose. In severe cases the speech is quite unintelligible, and the ingestion of food considerably impeded. The velum is absolutely immovable and does not respond to electric irritation; the pharyngeal reflex is abolished. Sometimes the palsy is unilateral (corresponding to the side where the exudate was heaviest ?); the uvula is drawn toward the sound side. In mild cases interference with speech is the only distinctly recognizable symptom. (2) When paralysis of the palate is severe, there is often associated partial paralysis of the deep pharyngeal, and of the laryngeal muscles; swallowing is greatly interfered with; there is tendency for the "food to get into the Sunday throat" (failure of the epiglottis to close); and contact of the food with the larynx incites a hoarse, spasmodic cough. The voice is weak or there may even be absolute aphonia. Paralysis of the laryngeal muscles (posticus paralysis) can be seen with the laryngoscope. (3) Paralysis of accommodation is not infrequent, although the phenomenon is not so noticeable in children. It shows itself in inability to do fine work or read, and the patient usually first consults the oculist. Subjective phenomena such as *muscæ volitantes* are rare. In older children inability to fix an object that is held near the eyes is readily recognized. I have also observed ocular palsies (abducens, oculomotor). (4) The patellar and tendo Achillis reflexes are almost regularly abolished early in the disease; in exceptional cases they may be preserved and even quite active (personal observation). In severe cases paresis of the legs, ataxia and inability to walk are observed. The legs are very much emaciated and the reactions of degeneration are present, but there is no pain either spontaneous or elicited by pressure on the nerve trunks. Other groups or muscles may become partially paralyzed, especially the muscles of the neck, as a result of which the head drops forward on the chest or is inclined to one side. Tremor, ataxia and paralysis may be present in the arms, and the abdominal and thoracic muscles are sometimes attacked in severe cases. Whether the cardiac weakness and sudden death from heart failure which occur in cases of severe diphtheritic palsy are due to the same causes as the nerve palsies is difficult to decide, for we must always reckon with the possibility of a direct infectious myocarditis. General depression, pronounced pallor and albuminuria are frequent concomitants of a severe palsy.

Postdiphtheritic neuritis presents many degrees of severity, from a simple peripheral paralysis with no more serious disturbance than nasal speech to severe general paralysis, and may be arrested at any of these stages. The way in which the paralytic phenomena make their appearance is usually as follows: paralysis of the palate, diminution of reflexes in the legs, then paralysis of accommodation, then paralysis of the neck and legs, of the larynx, and finally of the entire body. When the primary diphtheritic lesion is elsewhere in the throat, the paralysis begins in the muscles nearest the diseased focus instead of in the palate (abdominal paralysis after diphtheria of the umbilicus).

In most cases the disease lasts from 4 to 10 weeks and gradually ends in recovery. A duration of several months is possible. The dangers of the disease are heart weakness, inspiration pneumonia, paralysis of the diaphragm and general inanition. Hence the pulse, respiration and the urine (albuminuria is an unfavorable symptom) must be watched with the greatest care. Sudden death from heart failure may occur without warning.

According to the view which is generally accepted at the present time the pathologic basis of postdiphtheritic paralysis is an inflammation of various peripheral nerves. Positive changes in the spinal cord are particularly frequent in this form of polyneuritis and their significance has already been discussed in connection with that disease. They are degenerations of the anterior roots, changes in the cells of the anterior horns (Dejerine), increase in the neurogliar tissue, hæmorrhage into the substance of the spinal cord and into the spinal ganglia, and meningitic process (Oertel). In every instance we find disease of the peripheral nerves, either alone or in connection with the above-mentioned spinal changes (Preisz). The nerves exhibit degeneration of the parenchyma, inflammation and proliferation of the connective tissue, with new formation of cells and accumulation of leucocytes or fusiform swellings of the trunk of the nerve (P. Meyer). The cranial nerves also are frequently degenerated (Mendel, Oppenheim, Siemerling, Arnheim, etc.). The muscles also exhibit parenchymatous degeneration with increase of connective tissue (Hochhaus). In spite of the great variety of the changes observed, practically all the authorities are agreed that the essential pathologic change at the foundation of the clinical picture is inflammatory disease of the peripheral nervous system. According to Remak the beginning of the paralysis is found in the palate on account of the nerves of that organ being, so to speak, "immersed in the poisonous focus."

Experiments to produce diphtheritic palsy in animals artificially have not led to any definite results. According to Babonneix's attempt in this direction it appears probable that the diphtheritic poison is conveyed toward the brain along the nerves and not by the blood vessels.

The **diagnosis** in the presence of unmistakable evidences of an attack of diphtheria and beginning paralysis of the palate presents no difficulties. A doubt could arise only in cases of evident diphtheritic palsy without antecedent diphtheria. Experience leads us to believe that nondiphtheritic angina is also capable of producing paralysis of the palate. But cases of this kind are not altogether above criticism, and admit of two possible explanations: either that there may have been a diphtheria after all, or that the apparent angina was merely a symptom of some other grave disease which was the cause of a general polyneuritis. Hence, notwithstanding exceptional cases of this kind, it is a well-established fact, confirmed by an overwhelming number of cases, that a polyneuritis beginning with paralysis of the palate is practically always due to an antecedent diphtheria, and that the occurrence of the palsy may be the earliest evidence of the true nature of an apparently benign angina.

In most cases of postdiphtheritic palsy expectant and tonic **treatment** is all that is required; but whenever the disease has gone beyond the stage of simple paralysis of the palate, rest in bed with avoidance of all strain on the heart and regulation of the bowels is urgently indicated. If the paralysis interferes seriously with the taking of nourishment, artificial feeding with the stomach tube or nasal feeding may be necessary for a time. In any case the patient requires the most careful feeding. Sometimes it is found that fluids are regurgitated through the nose although the child is able to swallow semi-fluid and solid food. On the other hand, some children are able to swallow fluid or semi-fluid substances more easily than solid food. Artificial albuminous preparations, such as somatose, puro, sanatogen, etc., which are generally speaking superfluous articles, may be occasionally employed for the purpose of insuring a digestible and nutritious diet. Albuminuria may be disregarded in selecting a diet. When the patient is unable to take enough fluid, it is sometimes necessary to give small pieces of ice, to be dissolved slowly in the mouth, in order to relieve the thirst. If the nutrition is very much impaired, nutritive enemata may have to be employed.

Of internal remedies, strychnine, which was first warmly recommended by Henoch, is still the most useful. It is given hypodermically in doses of 0.0005 to 0.001 to 0.002 Gm. per diem ($\frac{1}{128}$—$\frac{1}{32}$ gr.). If there is any objection to giving strychnine hypodermically in children, tincture of nux vomica 2.0: 10.0 Gm. (30 gr.: 2½ dr.) (of bitter tincture) 5 drops (to a child of 3), 15 drops (to a child of 5) or 20 drops two or three times a day, to be followed by milk. Electricity should be tried in every case. If there is paralysis of the pharyngeal muscles, a local application of an electrode in the shape of a catheter (with a faradic current, or the stabile cathode), or stroking movements with the cathode applied to the throat, increasing the strength of the current until a deglutition movement is elicited. In paralysis of the extremities faradization or massage of the paralyzed muscles is recommended.

Recently injections of large doses of antitoxin, repeated several days in succession, have been administered in cases of postdiphtheritic palsy and, it is said, with very good results.

(*b*) *Alcoholic Neuritis*

In the child alcoholic intoxication manifests itself by cerebral, more often than by peripheral symptoms. In acute cases convulsions, irregularity of the respiration, delirium, coma or even sudden death are observed; in cases of chronic intoxication, increased irritability (as evidenced for example by excessive restlessness during the examination) insomnia, ill-temper, mental dulness, and epileptiform attacks.

Only a few cases of peripheral alcoholic neuritis in children are found in the literature (Lescynski, Jacob, Campbell, Zappert, etc.). In every case there is a history of long-continued indulgence in alcoholic beverages. It seems that beer, which can be taken for a long time in small amounts without producing any immediate symptoms, is particularly apt to cause peripheral neuritis.

The **symptoms** of alcoholic neuritis in the child are practically the same as in the adult. It is first noticed that the patient tires easily when walking, and this is soon followed by paresis of the legs and complete inability to walk. Ataxia is usually present and is most noticeable when a child which has been completely paralyzed in the leg begins to regain the power of walking. The deep reflexes of the legs are usually abolished, the reaction to electric stimuli is variable. Later in the course of the disease weakness of the arms and muscles of the trunk develops. An important symptom is pain along the nerve trunk, which the patient sometimes complains of spontaneously and which may be accompanied by hyperalgesia of the skin. The muscles also are sometimes sensitive on pressure. Œdema of the skin is occasionally observed. Other nervous disturbances, such as palsies of the ocular muscles, are rare in cases occurring during childhood. In children who are confined to bed on account of some other grave effect of alcoholic intoxication (heart, liver), the symptoms of polyneuritis may be quite overshadowed by the other symptoms present (Campbell).

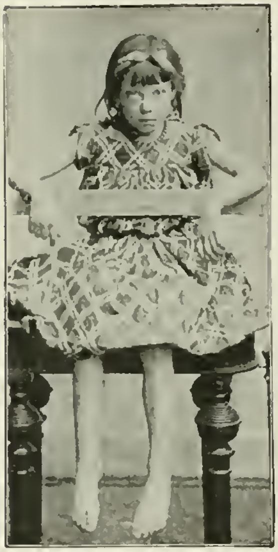
The **course** and **prognosis** in alcoholic neuritis are not bad unless the primary disease itself brings with it dangerous complications. Recovery is slow and may require many weeks or even months.

Something is known of the pathologic anatomy of alcoholic neuritis in children since one case (Campbell) ended fatally. The pathologic findings in the adult form the subject of a monograph by Heilbronner. With regard to the peripheral nerve changes, which predominate in the pathologic picture, and the spinal lesions, they have already been discussed in connection with polyneuritis.

It is evident that these cases should be a warning against giving alcohol to children; although, considering how rare they are compared with the frequent use of beer in childhood, the exploitation of these cases by the opponents of alcohol for purposes of agitation does not seem to be entirely justified. The *treatment* consists in withdrawing the alcohol, rest in bed, and faradization.

(c) *Lead Neuritis*

Fig. 55.



Lead neuritis. Paralysis of the extensors of the arms and feet. Probably hereditary.

Neuritis due to lead poisoning has been described more frequently in childhood than alcoholic neuritis (about 35 to 40 cases). The causes are exposure to lead in the workshops of lead workers, playing with toys or bits of silk containing lead, lead enamelled drinking vessels, swallowing various foreign bodies containing lead, ingestion of the metal in medicinal preparations, etc. The possibility of blood poisoning (of the cerebral variety) resulting from the use of diachylon ointment (Hahn) should be borne in mind, and we may also mention a strange case (reported by Anker) of hereditary lead poisoning in the child of a man who was suffering from that disease.

While referring the reader for the other symptoms of lead poisoning to the chapter on the intoxications, we shall here confine ourselves to a discussion of the nervous symptoms. These are somewhat different in children than in adults, the legs being regularly attacked by paralysis first, especially the peroneal muscles. Emaciation, reactions of degeneration, and abolition of the patellar and Achilles tendon reflexes are observed in the paralyzed legs. The arms are attacked later and then exhibit typical radial nerve palsy. As a rule there are no other disturbances to be found except dragging pains in the limbs. With regard to other symptoms of lead poisoning, extreme pallor and colic are usually present; but the blue line is not always found. Convulsions referable to lead poisoning of the brain have been observed in infants. Other nervous symptoms such as hemiplegia, tremor, spasm of the bladder, optic neuritis, disturbance of the eye muscles and disease of the cranial nerves, are extremely rare in children.

The **prognosis** of lead neuritis in children is not bad provided the cause is recognized and constant reintoxication with the metal can be avoided. If the cause cannot be removed, the disease may be greatly protracted. In cases in which the brain is affected, death with convulsions has been described, and in a case of Hahn's the brain was found to contain lead. But even in cases of peripheral neuritis a complete cure cannot always be promised. In one case under my observation there has been for the past two years paralysis of the peroneal nerve. Relapses are by no means rare when external conditions are unfavorable.

The *pathologic anatomy* is the same as in alcoholic neuritis.

(d) *Arsenical Neuritis*

After the ingestion of a toxic dose of arsenic, and especially after a protracted course of medicinal doses of Fowler's solution as, for example, in chorea, poisoning has not infrequently been observed with certain nervous features which may be briefly described.

The **symptoms** are those of polyneuritis; pain in the distal portions of the extremities and paralyses occupying the foreground in the clinical picture. The paralysis is accompanied by atrophy and the presence of the reactions of degeneration in the muscles; the lower extremities, particularly the legs, are chiefly affected. When the arms are attacked, atrophic paralysis of the muscles of the hand often results. Sometimes ataxia is more pronounced than paralysis (pseudotabes). The patellar reflexes are usually abolished. Painful points are found here and there along the nerve trunk and objective disturbances of sensation are often observed. Trophic disturbances of the skin in the form of hyperidrosis, glossy skin and pigmentations are not infrequent. The occurrence of herpes cannot be regarded as a component of the picture of arsenical polyneuritis, since it may represent the only sequel of arsenic poisoning.

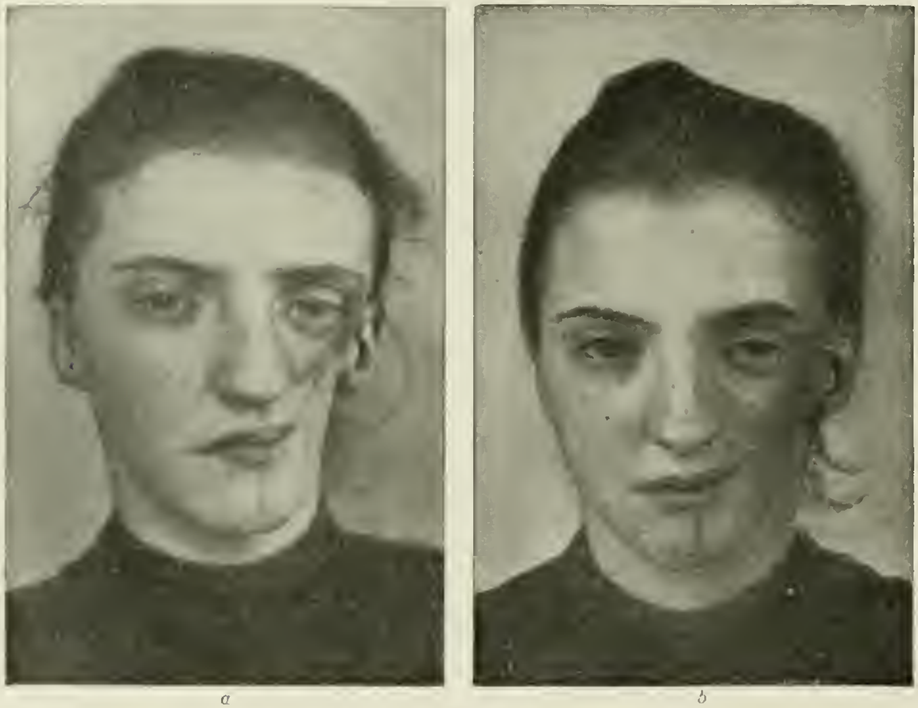
The **course** is usually satisfactory. Oppenheim reports a few cases of contractures in the paralyzed extremities.

V. HEMIATROPHY OF THE FACE

Hemiatrophy of the face is a progressive emaciation of one-half of the face including the skin, muscles and bones. The disease is not very rare in childhood, at least the beginning of the malady can in many cases be traced to that period of life. After the thirtieth year the disease according to Möbius does not occur. Girls are more frequently attacked than boys and the left side more frequently than the right. In rare cases both sides of the face are affected by the atrophy (as observed by the writer in a girl of seventeen).

The **cause** of the disease is unknown, as its nature has as yet never been properly explained. The slight knowledge we have of the pathology of the disease (Mendel, and especially Löbel and Wiesel) would seem to indicate an interstitial inflammatory process in the trigeminal nerve including the Gasserian ganglion as the original cause of the disease. According to this view hemiatrophy of the face is therefore a chronic inflammatory disease of the peripheral portions of the trigeminus. It is possible that inflammatory processes in the various portions of the head (erysipelas, angina and the like), mention of which is sometimes found in the history as forerunners of the disease, really have some etiologic significance. In other cases there is a possibility of toxic or infectious substances having invaded the tissues through the tonsils and produced a hemiatrophy of the face (Möbius).

Fig. 56.



Hemiatrophy of the face. (*a*) Before paraffine treatment. (*b*) During paraffine treatment. The treatment is to be continued.

The disease begins with atrophy of a limited portion of the skin of the face, which becomes attenuated and loses its subcutaneous fat, so that it can be taken up in minute folds. Sometimes brownish discoloration of a small portion of the skin is observed. These changes usually begin in the cheek, in the canine fossa. The atrophy rapidly spreads to the muscles and bones, causing depression especially of the zygoma and of the upper jaw. The atrophy ultimately effects the entire half of the face, which presents a sharp contrast to the healthy side, particularly along the median line, the forehead, lips and chin. The line of separation is sometimes convex toward the sound side, as though the latter were endeavoring to surround the diseased half. The tongue, the upper and lower jaws, and the pharyngeal structures share in the hemiatrophy in severe cases, and the hair falls out on the affected side of the scalp. Neuralgic pains are sometimes complained of at the beginning of the disease as toothache. Paralysis of the muscles of the face is entirely absent and, although the muscles of mastication share in the general emaciation, they present no functional weakness.

The **course** of hemiatrophy of the face is progressive in so far as in the majority of the cases the entire half of the face is ultimately affected. The disease then becomes arrested and it is even said that in some cases the diseased half of the face fills out again. In some instances the atrophy is confined to a small portion of the cheek.

The **diagnosis** is clear at the first glance. The only possible source of error is an old facial palsy with secondary asymmetry of the face simulating hemiatrophy. In fact, Fromhold-Treu in his monograph of this disease mentions a large number of alleged cases of hemiatrophy which do not belong to the group at all.

The **treatment** of circumscribed hemiatrophy of the face is powerless and we therefore hail with joy the recent efforts of Gersuny and Moskowicz to correct the deformity by repeated subcutaneous injections of paraffine in the affected area. The result is remarkably good, and as the disease, while painless, is nevertheless greatly disabling on account of the striking change in the expression of the face, the results so far as the patient is concerned are equivalent to an actual cure of the disease.

VI. NEOPLASMS OF THE PERIPHERAL NERVES

The extension of neoplasms to peripheral nerves and the occurrence of nodules (neuromata) within the nerves are subjects that chiefly interest the surgeon and are of no importance in children's practice. Similarly the rare occurrence of multiple painful nodules in the nerves (tubercula dolorosa) and of a congenital plexiform neuroma of the trigeminus are without significance to the pediatrist. On the other hand, general neurofibromatosis or Recklinghausen's disease which, although extremely rare, has been observed in the child (Berggrün) and which probably depends on congenital predisposition, deserves brief mention in a work on pediatrics. The disease may be a hereditary or rather a family one, and for this reason may be included among the above-described endogenous diseases.

The disease consists in the appearance of numerous nodules and pigmented patches in the skin and in tumors of the nerve trunks. Sometimes, though not always, there is spontaneous pain or pain on pressure in the skin, and the muscles are sensitive; quite often the disease runs its course without producing any subjective symptoms.

The gravity of the affection may be much increased if the nerve tumors make their appearance in the roots of the spinal and cranial nerves. The symptoms are quite atypical and difficult to interpret and may produce a clinical picture resembling that of tumor of the spinal cord or brain, and lead to a fatal termination. The point of exit of the auditory nerve from the base of the brain is a favorite localization for this type of brain tumor, which sometimes is solitary (neurofibroma of the auditory nerve). When none of the cranial nerves are involved, the disease runs a slowly progressive course, although temporary remission and even involution of the tumor have been observed.

SECTION X.

DISEASES OF THE MUSCULAR APPARATUS

(Congenital absence of muscles, myositis)

The pathology of the muscular system is even to-day still a stepchild of internal medicine. In the case of muscles more than other organs we are very much inclined to regard any disease as an accompaniment or sequel of some general pathologic condition, or to classify such diseases with other clinical conditions which they may resemble. If we were to accord to the diseases of the muscular apparatus the autonomy which they deserve, we should have to mention in this place so-called rheumatic myalgia, rachitic muscular relaxation, as well as the muscular dystrophies which have already been discussed, Thomsen's myotonia, Oppenheim's myotonia and possibly also myoplegia, myasthenia and many forms of pseudoparesis occurring in childhood. But aside from the limitation of space, it is not the object of this work to inaugurate changes in the customary classification of pediatric diseases, and the diseases of the muscular system have accordingly been treated in other sections of the book, so that now we have left to discuss only congenital absence of muscles and inflammations of muscles.

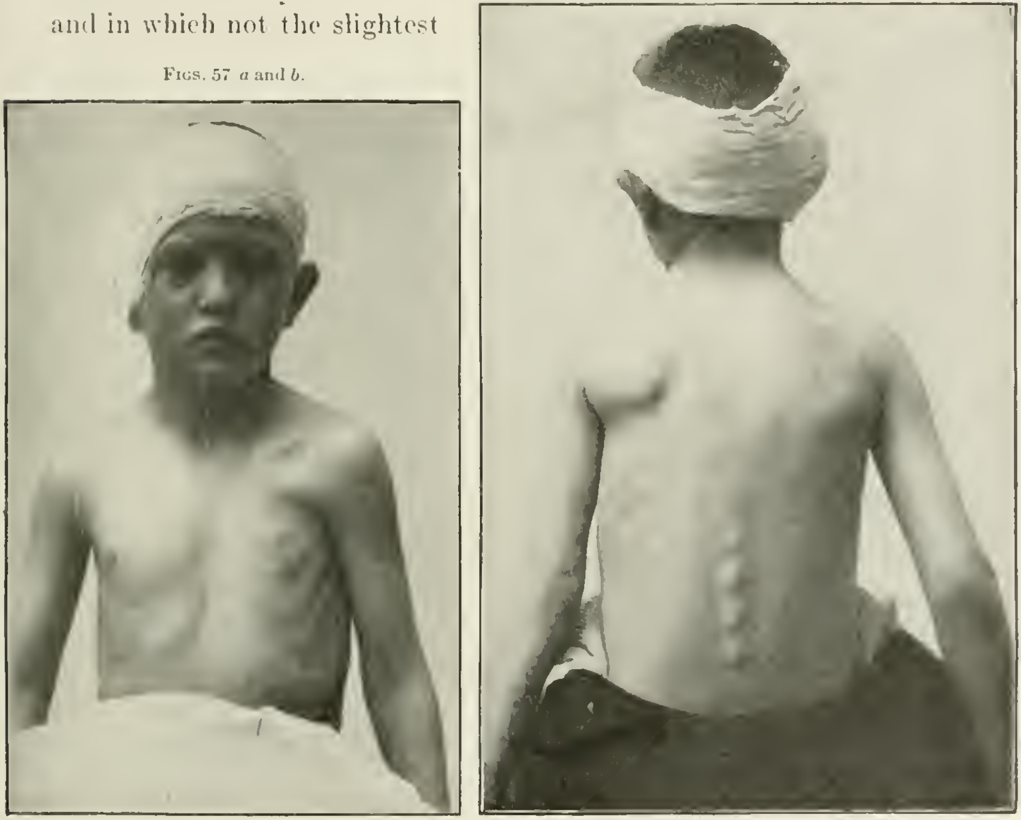
CONGENITAL ABSENCE OF MUSCLES

It is a matter of surprise that the reported cases of congenital absence of muscles have to deal much less with children than with half-grown or adult individuals. The reason probably is that the anomaly does not cause any marked symptoms and therefore does not become apparent until the muscles of the body are fully developed or some secondary phenomenon (such as an underdeveloped breast in absence of the pectoral muscle) calls attention to the condition. Moreover, otherwise healthy individuals do not, as a rule, undergo medical examination until they are drafted for the army or apply for life insurance. Now that school inspection is becoming general, it is probable that congenital absence of muscles will be reported among school children.

Absence of the pectoral muscles is the most frequent anomaly; deficiency of the trapezius, serratus magnus, quadriceps femoris and other muscles is not so common (see Bing's statistics). With reference to congenital absence of the ocular muscles see infantile nuclear atrophy.

The defect may be complete or partial, a point to be decided by post-mortem examination, which reveals various grades from complete absence of the muscle to the presence of numerous rudimentary muscle fibres. That in a number of cases at least the condition is due to a congenital aplasia and not to disease acquired during intra-uterine life is shown by a case of absence of the pectoral muscle in a child five days old examined by Rückert and in which not the slightest trace of inflammation could be found in the remaining muscle fibres.

Figs. 57 *a* and *b*.



Congenital muscular defects. Absence of pectoral muscle, deformity of the thoracic wall. The nipple is indistinct and the scapula higher on the affected than on the normal side. (The bandage on the head has nothing to do with the case).

The most typical picture is seen in congenital absence of the pectoral muscles. The normal fulness of the anterior chest wall is replaced by a depression, and no muscular tissue can be felt. Quite often other pathologic conditions are associated with this defect, such as abnormalities of the thorax, absence of one or two ribs, imperfect development and webbing of the fingers of the hand of the same side, atrophy of the skin, deficiency in pigmentation around the nipple, which is higher than on the other side, and imperfect development of the breast. Steche recently attempted to establish a special type of muscular deficiency in the coincidence of all these symptoms. Congenital upward displacement of the shoulder (Sprengel) is another condition that often accompanies atrophy of the pectoral muscles.

The appearances in cases of absence of other muscles are less pronounced and are practically the same as in acquired paralysis of the same muscles.

Although the function of the muscle in congenital aplasia is obviously lost, the actual disability, so far as the patients are concerned, is relatively slight. They learn early in life to make as much use as possible of other, sound muscles. Thus, even in the absence of the pectoral muscles, the boy often learns to do gymnastics, to swim, etc., without any trouble. Complicated movements of the arms, however, are performed awkwardly and without the proper degree of strength, so that a man with absence of the pectoral muscles is regarded as unfit for military service.

Several muscles may be absent in the same individual, but the defects are never symmetrical on the two sides. This is an important point in the differential diagnosis between congenital absence and acquired muscular atrophy, which also differs from the former by the mode of onset and the progress of the disease.

Muscular defects are permanent and no treatment is possible. If the disability is marked, surgical relief by transplantation of tendons or muscles may be considered.

INFLAMMATIONS OF MUSCLES (MYOSITIS)

We distinguish local and general, purulent and nonpurulent inflammations of muscles.

Local purulent myositis results either from trauma or from inflammation of neighboring organs, or from some general infection. Multiple purulent myostitis is a rare complication of universal sepsis, scarlatina and other infections. Nonpurulent inflammation of individual muscles may result from rheumatism, scarlatina, gonorrhœa, typhoid fever, etc., or from injury. It is possible that syphilis (Hochsinger) and tuberculosis also lead to inflammation of individual muscles without abscess formation.

These different forms of myositis are all part of other diseases and have received due attention in the appropriate places.

Polymyositis or non-purulent inflammation of the entire muscular system is an independent disease and often quite difficult to recognize. It occurs either as a primary affection without any recognizable cause or as a sequel of some parasitic infection, particularly trichinosis. In primary polymyositis the intestinal symptoms are at first so pronounced as to suggest that the causative organism first effects an entrance through the intestinal tract.

Lorenz has subdivided primary polymyositis into the following groups: dermatomyositis, hæmorrhagic myositis, myositis with erythema multiforme, and fibrous myositis. Certain other less pronounced forms of polymyositis might be added. These different varieties of multiple myositis, however, do not represent so many different clinical types, the classification being based on individual symptoms, which may characterize the clinical picture of myositis as a whole either clinically or pathologically. Myositis following a polyneuritis is designated neuromyositis. Primary progressive ossifying myositis is a special disease which will be discussed separately.

The characteristic features of polymyositis are briefly as follows:

The prodromal stage, lasting several days, is marked by general malaise with fever, anorexia, vomiting, pain in the limbs and headache; the fever gradually rises; the patient's subjective state rapidly becomes worse; and albuminuria sometimes makes its appearance. Œdema then develops in the eyelids and in the face and, as a rule, spreads rapidly to the surface of the entire body. At the same time the muscles of the face become rigid, boardlike and very painful. The fever continuing, sometimes increasing by abrupt rises, the myositis spreads to the other portions of the body, particularly the extremities; the hands and feet being as a rule less severely damaged than any other portions. The muscles feel swollen, hard and doughy, and are extremely painful. The deep reflexes are usually diminished; the skin reflexes, as a rule, persist.

The patients are quite unable to move. The general condition is greatly impaired by the pyrexia, which suggests that of typhoid fever, by the pain, and by the difficulty of taking nourishment. The course may be quite rapid and death may result in a short time from involvement of the muscles of respiration, the heart and muscles of deglutition. In favorable cases the œdema, fever and gradually also the muscular swelling subside, and the patient recovers within a few weeks or months. Sometimes the course is subacute or even chronic and interrupted by exacerbations.

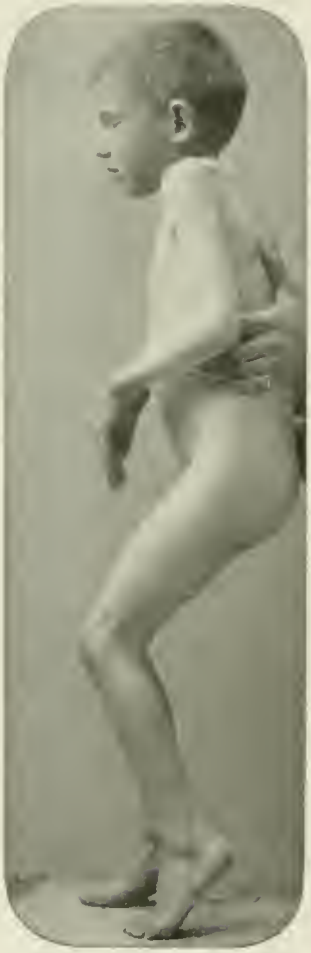
The **pathology** consists in acute inflammation of the muscle parenchyma and of the interstitial tissue. When the inflammatory cutaneous œdema is pronounced, the term dermatomyositis is used (Unverricht). Hæmorrhagic polymyositis is accompanied by a hæmorrhagic exudate and other hæmorrhages into the skin and mucous membranes. The heart is usually attacked in this form, and the prospect of a favorable outcome is very slender. When the inflammatory process exhibits a more chronic character from the beginning and is attended by proliferation of connective tissue in the muscle, the disease is called *fibrous myositis.*

Polymyositis usually runs a less violent course in children than in adults. The cases collected by Schüller all ended in recovery [Janicke, Schultze, Köster, Cassirer, Oppenheim (the last case is doubtful)]. The case from my own clinic, which was described by Schüller, when at its height bore a great similarity to a form of cerebral infantile palsy designated "cerebral rigidity." In this case the course was mild, but in other respects like the above-described typical clinical picture.

Cases of myositis are susceptible only to purely symptomatic *treatment.*

Trichinous polymyositis, the cause of which is the entrance of the embryonal parasites into the intestine and their migration by way of the lymph or blood channels into the muscular capillaries, with the production of irritative symptoms is quite similar to primary myositis. Accordingly, trichinosis myositis differs from the primary form only by its cause, and the clinical picture in the two diseases may be clinically so nearly identical that the differential diagnosis must be based on etiologic and other extraneous factors. The occurrence of similar cases in the same locality or in the same family, the prominence of the primary intestinal symptoms, the presence of the parasites in the feces, eosinophilia and possibly the demonstration by means of the X-ray of numerous calcified trichina capsules in the thinner muscles (Gocht, Schüller) are points in favor of trichinosis. The course of trichinosis is also milder in the child than in the adult, recovery frequently resulting by calcification of the incapsulated parasites. Other parasites (echinococcus, cysticercus, sporozoa) may produce similar diseases of the muscles.

Fig. 58.



Acute polymyositis in a boy of seven. The contractures resemble those seen in cerebral rigidity.

OSSIFYING MYOSITIS

Localized bone formation in the muscles results from constant irritation of certain muscles ("rider's bone, Exerzierknochen"), more rarely from injuries to the muscle. This ossifying form of myositis has practically no significance in pediatrics.

On the other hand, progressive multiple ossifying myositis is a disease that is peculiar to childhood. Of 51 cases collected by Lorenz, 11 occurred in the first year of life, 16 between the ages of 1 and 5, 11 between 5 and 15, and only 7 among individuals more than 15 years of age.

This remarkable incidence must be taken into account in formulating theories about the pathogenesis of this form of myositis, and points strongly to some inherited or congenital injury as the cause of the disease. This, however, is not the case, since the disease is not so far as we know, either hereditary or family, but always develops gradually. Nor can it be denied that in many cases the exciting cause appears to be an injury, albeit one which otherwise would be disregarded in childhood. We are accordingly forced to assume a constitutional anomaly (Münchmeyer) that renders the muscular tissue abnormally sensitive to external irritation. The affection is therefore not an endogenous disease according to the definition we have given, but represents a deviation from the normal in the tissues of the affected children. That these children are actually abnormal from birth is shown from the frequent combination of ossifying myositis with smallness of the large toes and of the thumbs (Gerber).

Clinically we observe local signs of inflammation in certain muscles following injury accompanied by fever, pain, swelling, and œdema. These symptoms subside and are followed by the development of a doughy, muscular wheal, which may persist for years without change. Similar alterations develop again and again in various muscles in connection with external injuries. Sometimes the disease begins in a number of muscles without any known cause, and in such cases the nodular foci of inflammation may temporarily disappear. Gradually we are able to see or feel small bony kernels within these nodes, which increase in size and ultimately involve large portions of the muscular apparatus. Pain is usually slight, but the interference with movements steadily increases and is a great annoyance to the patients.

The muscles of the neck and back are usually attacked first, then the muscles of the extremities and finally the masseter and temporal muscles, greatly interfering both with locomotion and mastication. While at first no more than a certain awkwardness is noticed in the child's movements, and it cannot stand erect, there gradually develops great interference with every kind of movement or even complete loss of mobility. The entire body is bent until finally the patients become entirely helpless and have to keep their beds.

The **progress** of the disease takes place by successive stages, interrupted by long intervals during which the disease is arrested. Years elapse before the malady reaches its height, so that well-marked cases are more frequently seen in adults than in children.

Pathologically we recognize a stage of acute myositis followed by proliferation of intramuscular connective tissue (fibrous myositis), and true ossification in the muscles. Ossification starts partly from the bone itself in the form of exostoses, hardening of the muscular attachments and bony unions between different bones, and partly within the intramuscular connective tissue or even within the muscle fibrils. The muscle elements atrophy and are replaced first by proliferated connective tissue and later by bone substance. The ossification does not involve the muscles in their entire extent, but occurs rather in the form of disseminated nodules within the muscle tissue.

The **prognosis** is grave, not only as regards recovery but also, in severe cases, with regard to life on account of the interference with respiration and the difficulty of administering nourishment.

The **diagonsis**, which presents no difficulties in the terminal stage, is practically impossible at the beginning, and the disease is at first usually mistaken for some rheumatic affection.

The **treatment** is hopeless. At best, the progress of the disease may be slightly retarded by avoiding injuries, and by means of baths, and the iodides administered by inunction.

The above description applies only to the typical form of progressive ossifying myositis. The many deviations and unusual varieties which we are forced to ignore in the present discussion merely serve to accentuate the enigmatical character of this interesting disease.

FUNCTIONAL DISEASES OF THE NERVOUS SYSTEM

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TRANSLATED BY

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INTRODUCTION

THE term functional is applied to all diseases the pathologic basis of which is still unknown, and which on that account we contrast in a certain sense with the previously described organic diseases. If this principle is kept in mind, it will be readily seen why heterogeneous symptom-complexes, due to a variety of etiologic factors, are included in the same group of functional nervous diseases, and that the dividing line between functional and organic affections changes from time to time as our knowledge of pathogenesis increases, so that at the present day the classification of certain disease-types, such, for example, as chorea and epilepsy, among the functional diseases meets with objections which are not altogether unjustified.

No one can doubt that for every functional disturbance there must be a corresponding anatomical change in the nervous system; but, in accordance with the fugaciousness of the symptoms, the pathologic lesions are in some way peculiar, and as yet we have no conception of what this peculiarity consists in.

Aside from the pathologic viewpoint, from which we approached the subject in the beginning, functional diseases also possess certain *common clinical characteristics* which justify their classification in one large group. The most important characteristic, which is common to almost all functional diseases and on which the others appear to depend, is that the symptom-complexes depend on a certain constitutional anomaly of the entire nervous system. This does not prevent the fact that certain definite symptoms—in harmony with the broad principle of localization—do not have their seat in some unknown anomaly of a definite portion of the nervous system; but if we carefully examine the entire individual, both from the neurologic and the psychic standpoint, and if we take his subsequent development into consideration, we will discover phenomena which cannot be explained on the ground of a single circumscribed localization of the morbid process. This will be explained more fully when we come to describe the neuroses, such as hysteria and neurasthenia.

If we follow out this conception to its logical conclusion, we will understand why the symptoms that are observed exhibit such a great variety in themselves, and vary to such a remarkable degree in the same individual. This will also be explained later by means of illustrative cases. It also affords an explanation of the fact that heredity plays a much greater part in the functional than in the organic nervous diseases. Thus not only direct similar inheritance from both parents is observed in one or more of their children, as well as the family incidence of certain nervous diseases; but, on the other hand, we frequently come across cases of dissimilar inheritance which challenge our attention. Finally, we are able to understand that the prognosis of a single symptom is not identical with the prognosis of the disease as a whole, and that we must be prepared for the sudden disappearance of one symptom and the equally sudden appearance of another that may be quite different externally, although sprung from the same pathologic soil.

Any classification of functional nervous diseases, composed as they are of heterogeneous things, is necessarily more or less arbitrary. It seems to us wisest to erect two main groups. The first contains those diseases in which somatic symptoms are produced by anomalies in the psychic life of the individual, in which, as Wernicke has explained in the case of the psychoses, the association-system is the seat of the disease. The type of this group is hysteria. The second group embraces all those functional disturbances in which we may expect to find a primary lesion of the projection-system. The representative of this group is chorea. Neurasthenia occupies an intermediate position between the two groups. Its symptomatology is much more pronounced than is the case in diseases belonging to the two groups mentioned, being composed of psychogenic or psychic and primary somatic disturbances.

Since the psychic development is much less advanced in children than in adults, the second group, that of the primary somatic disturbances, interests us chiefly in this connection, particularly as these disturbances are the earliest that occur in childhood.

We shall accordingly begin our systematic exposition of the subject with the large symptom-group of the convulsive diseases.

CONVULSIVE DISEASES

The fact that children in general, and particularly those in the first two years of life, are much more frequently attacked by convulsions than adults long ago arrested the attention of physicians and gave rise to many theories for its explanation. It does not appear necessary to enumerate all these theories and we shall therefore mention only the following:

The cause of the frequent occurrence of convulsions was assumed to reside either in the peculiarities of the diseases to which children are subject, and which differ from the diseases of adults, or in peculiarities of the childish organism or nervous system; or finally, in both these pathologic and physiologic factors.*

The most important period in the *history of infantile convulsions* is marked by the promulgation of Soltmann's hypothesis. This observer found by experimental investigation that in newborn dogs, cats, and rabbits the motor cortical areas discovered by Fritsch and Hitzig cannot be excited electrically, and are probably incapable of functionating. He concluded therefore that they are incapable of exercising either an innervating or an inhibiting influence on subcortical motor centres. In order to apply these discoveries to the human newborn infant there suggested itself a method which promised success, namely, a comparison of the medullary striation in the animals experimented upon and in human infants. Soltmann's investigations in this direction, which harmonize with similar ones made by other observers, showed that the human infant requires from twelve to eighteen months to attain the stage of development of an animal from ten to twelve days old, in which irritation of the cortex already produces movements on the opposite side of the body. From this Soltmann concludes that the inhibiting function is not developed, and does not become effective before that period in man. These results appeared to explain the frequency of convulsions in infants under eighteen months of age. Another interesting result of these animal experiments is that the nerves of newborn animals are much less irritable, and that even a small number of single electric stimuli in the second produces tetanus in a nerve-muscle preparation taken from a newborn animal, while under the same conditions in the adult animal each individual stimulus elicits a single contraction which can be distinctly separated from every other. In harmony with this phenomenon Soltmann found that the myogram of the single contraction in the newborn is flatter and that the contraction is more sluggish than in the adult animal.

Investigations by the same author to determine the time at which these abnormal conditions in the newborn change to the conditions as we know them in the adult, yielded a further noteworthy result, which appeared to be calculated still further to elucidate the frequent occurrence of convulsions during the later part of infancy. It was found that the irritability of the peripheral nerves attained the maximum, or even exceeded the maximum for a later period of life at a more rapid rate than that of the full development of the inhibitory centres.

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* It is needless to say we omit from the present discussion convulsions [illegible] in childhood as the result of exogenous intoxication. A table of intoxications which are said to be [illegible] of [illegible] convulsions in children will be found in an article by Hochsinger, Deutsche K[illegible], Vol. VII, [illegible].

"At about this time," Soltmann writes in speaking of the period between the fifth and the ninth months of life, "the irritability of the peripheral nerves is already quite considerable, perhaps even greater than in the adult; while conversely the mechanism of inhibition and the volitional faculties (the psychomotor cortical centres), although they have begun to develop, are by no means sufficiently powerful or sufficiently definite in their action to offer an efficient bar to the ready transmission of reflexes. This explains," Soltmann goes on to say, "that a quite insignificant irritation affecting the infant during this period of life, even if it does not appear to exceed the bounds of the physiologic, as, for example, the eruption of a tooth, which at another time would not produce any disturbance of any kind, is quite sufficient to bring on a convulsion."

The objections that were raised against Soltmann's doctrine of "physiologic spasmophilia" or "increased disposition to reflex irritation" in infancy and their untenability will be referred to again later.

Of the remaining theories in regard to the pathogenesis of functional convulsions in childhood we shall briefly mention the three which, in our opinion, are the most important and for the present shall not indulge in any critical comment, which will develop naturally in the course of our exposition of the subject.

The first may be briefly designated the *autointoxication hypothesis.* Bacterial toxines, on the one hand, and poisonous substances resulting from bacterial decomposition of the intestinal contents such as the diamins, on the other, have been mentioned in this connection. Later acetone, ammonia, carbaminic acid and similar substances, which enter the blood when the antitoxic function of the liver is insufficient, were credited with the power of producing the convulsions. Under certain conditions the sudden access of fever, overloading of the blood with carbon dioxide, and disturbance of the osmotic relations between the blood and the tissues were also believed to play an etiologic part in the production of convulsions.

The second theory is the one advocated by Kassowitz. According to this theory convulsions—and a few other nervous diseases—in infancy, represent the concomitant symptoms or sequelæ of *rachitis.* They are supposed to be due to circulatory disturbances in the cerebral cortex, which in turn depend in some way on the hyperæmia of the cranial bones in rachitis.

The third and last of the theories that we shall mention here was advanced by Baumé as early as 1805, and in our own times chiefly defended by Féré. According to this theory infantile convulsions are merely a *special form of epilepsy* peculiar to the age of the affected individuals and characterized by a more favorable prognosis.

It appears from the foregoing that Soltmann made the best attempt to explain the undeniable fact that functional convulsions are much more frequent during early childhood than at any other period of life.

But by this time we have outgrown his ingenious theory. Its downfall is not due to the fact that later investigations by Tarchanoff, Lemoine, Paneth and others have shaken its experimental foundations; nor to any change in our views with regard to the relations existing between functional power and electrical irritability in a nerve organ; nor to this or that secondary objection. His theory was shipwrecked on the contradiction which is found to exist between his assumptions and the results of clinical observation. Fleischmann was one of the earliest to object that his theories could not be brought into harmony with clinical experience, because in reality even the most intense stimuli, such as burns, the actual cautery, intestinal ulcers, peritonitis, etc. by no means often elicit convulsions.

Soltmann himself was quite aware of this weak point and later assumed, in addition to the *causa physiologica interna, i.e.*, the increased susceptibility to reflex irritation, and the *causa pathologica externa, i.e.*, the irritation which produces the convulsions, a *causa pathologica interna*, without however associating even a hypothetical conception with this term, although such an internal pathologic cause would alone explain the individual spasmophilia of certain children.

Not until quite recently have any additional facts or possible theories been brought forward in explanation of this point. Individual pathologic spasmophilia, which Thiemich in 1899 mentioned in his literary review on Convulsions in Childhood, is no longer a vague expression intended to hide our ignorance, but has come to signify an exact clinical finding. It is characterized by exaggerated mechanical and electrical irritability of the peripheral nervous system before and after the convulsions, and in the interval of freedom between the attacks. Children of this type are peculiar in the behavior of their peripheral nerves, so that we are justified in speaking of a special nervous state, which hitherto has usually been termed a tetanoid condition. Historically the latter term owes its origin to the fact that the anomaly which is a peculiar feature of the condition was first discovered in tetany; but it is too narrow, and has already led to misunderstandings (Hochsinger). We shall therefore make use of the more comprehensive term "spasmophilic condition" (Heubner) "or spasmophile diathesis" (Finkelstein).

By the term *spasmophile diathesis* of infants we mean a constitutional anomaly which is recognized by a mensurable mechanical and electrical overexcitability of the nervous system, and which produces a pathologic predisposition to certain partial and general clonic and tonic convulsions.

Accordingly, a number of *convulsive diseases*, which we shall presently have to describe, are considered together as being by virtue of this abnormal reaction of an identical type and only different manifestations of the pathologic spasmophilia of early infancy.

The exaggerated irritability which constitutes the peculiarity of the spasmophile diathesis, can in marked cases be determined by the presence of increased mechanical irritability in one or several peripheral nerves. Tapping the nerves with the percussion hammer at accessible points, such, for example, as the well-known nerve points of Erb, elicits a short and more or less violent contraction in the region which these nerves supply with motor impulses. The phenomenon is most clearly seen in the facial nerve, tapping of which produces unilateral contraction of the muscles of the face—the facial phenomenon or so-called Chvosteck phenomenon—which will be discussed again later in more detail.

It requires not a little experience on the part of the investigator to determine the effect, *i.e.*, the amplitude of the contraction produced by the stimulation; and one is also in danger of being led into error when the mechanical irritability of the muscle is increased. The latter has no pathognomonic significance whatever, as it also occurs in various conditions of increased irritability as, for example, in neurasthenia and the various cachexias.

As no clinical method has as yet been devised for measuring the mechanical irritability of a nerve, electrical examination is of much greater practical value. Erb established an increased faradic and galvanic irritability in the tetany of adults. After this had been confirmed in the case of children by Burckhardt, Kalischer, Escherich, Ganghofner and Hauser, Mann and Thiemich carried out a series of comparative investigations on a large number of children, some healthy and some suffering from tetany, which yielded a typical law of contraction, and thus rendered possible a still more delicate differentiation of normal and pathologic findings. The results may be tabulated as follows:

	ClC	ClC	OC	OC
Average findings for normal children	1.11	2.21	3.63	8.22
Manifest tetany or spasmophile diathesis	0.63	1.11	0.55	1.94
Latent tetany or spasmophile diathesis	0.70	1.15	0.95	2.23
Previous tetany or spasmophile diathesis	1.83	1.72	2.3	>7.9

The figures in this table were obtained by examining the median nerve at its most irritable point in the bend of the elbow, and the values are given in milliamperes. The children were never anæsthetized. The indifferent electrode, 50 square cm. in size, was placed on the chest, and a Stintzing normal electrode, 3 square cm. in size, was used for the active electrode. It should also be mentioned that the normal children were all more than eight weeks of age, because below this age the irritability of the peripheral nerves is much less (Westphal, Mann). The tetany cases were all more than eight weeks old.

A glance at the average figures collected in the table shows the greater irritability of children suffering from manifest or latent tetany, and the return to the normal immediately after the subsidence of the disease; but owing to the wide individual variations, which do not appear in the average figures, it is important to take account of the extreme values also. While referring for details to the above-mentioned thesis of Thiemich, we may mention the following diagnostic points: In tetany (the spasmophile diathesis in general) the values for ClC are for the most part lower than in normal children, although they may reach the normal level or even exceed it. The almost regular preponderance of AnOC over AnClC in tetany is of importance, as it very rarely occurs under normal conditions. *The criterion, however, is the behavior of KOC. Values below 5.0 m.a. must be regarded as pathologic, while values above 5.0 m.a. are normal:* Testing for KClT is uncertain in children without anæsthesia and cannot be substituted for KOC; nor is faradic examination of greater value.

These abnormalities in the reaction of the nerve to the irritation of the galvanic current, which can be determined by anyone possessing a little practice and dexterity, constitute the chief characteristic of the spasmophile diathesis.

This anomaly is the basis of the clinical pictures with convulsions of tetany, eclampsia, laryngospasm and apnœa, with which we have long been familiar, and also stands in close etiologic relationship to some at least of the cases of sudden death in early infancy without adequate anatomical findings.

CLINICAL FORMS OF SPASMOPHILIA

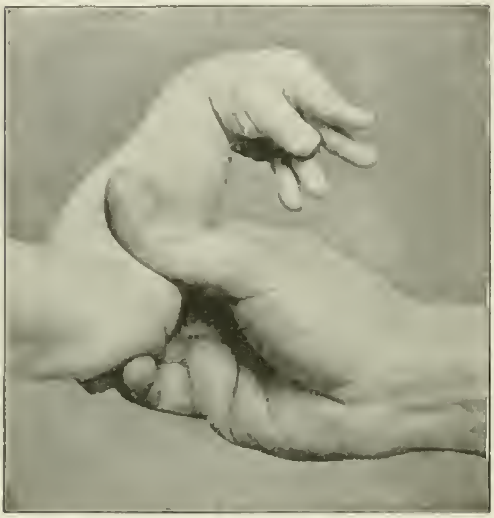
(*a*) TETANY

For the history of this affection the reader is referred to Frankl-Hochwart's exhaustive article on the subject in Nothnagel's Encyclopedia (Handbuch).

The chief **symptom** of the manifest form consists in tonic convulsions of the extremities, which are frequently accompanied by paræsthesia in the affected limbs, while consciousness is always preserved. The convulsions always occur in the upper extremities and force the hands into what is known as the "obstetrical position," which is well shown in the accompanying figure (Fig. 59). If, as is usually the case, the arms are flexed on the trunk, and the forearms and hands held in a position of flexion (the picture of "Pfötchenstellung"), the position assumed by a dog when he is "begging" (see Fig. 61) is produced. The lower extremities do not always share in the convulsions; if they do, they are usually flexed at the hip and knee, while the feet are in a position of varus or equinovarus with pes cavus contracture, as illustrated in Figs. 60 and 61.

The convulsions appear suddenly and last several hours or even days, to reappear after remissions of equal duration; or they may disappear altogether. In well-marked cases, in which œdema gradually develops in the dorsum of the hands and feet, these latter are rigidly fixed, and every attempt to correct their position elicits a cry of pain. In milder cases the attitude is the same during rest, but the resistance offered to passive movements is slight, and by exerting the will-power the children are able to overcome the forced position from time to time, long enough to use their hands for grasping things. During rest the hands return to the obstetrical position. The same relaxation of the tonus is observed toward the end of an attack of tetany.

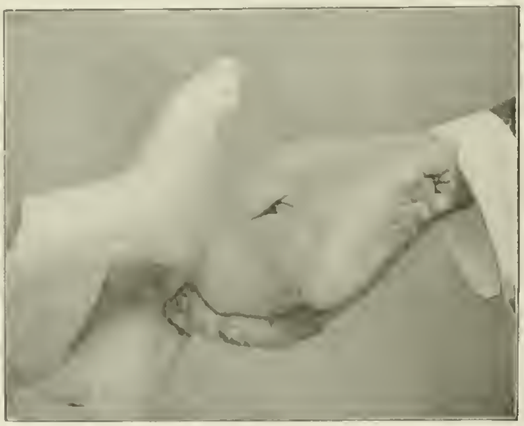
Fig. 59.



Typical tetany position of the hand. A child twelve months old.

During a severe attack the tonic contracture may spread to the muscles of the trunk and face, producing a rigid expression of countenance, with wrinkled brow and mouth protruded like a snout (carp-mouth). Among rarer events are retention of urine from spasm of the sphincter of the bladder, disturbances of deglutition, pupillary rigidity and dyspnœa, all of which have been described; all these phenomena are regularly associated with the typical attitude of the extremities.

Fig. 60.

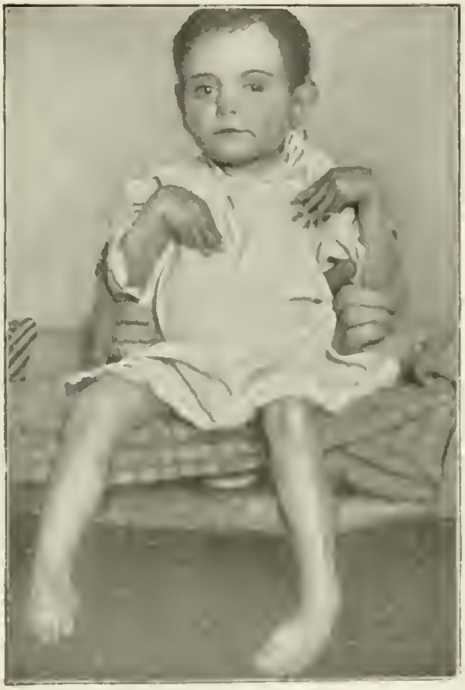


Typical tetany position of the feet. The same child as shown in Fig. 59.

Although the position of the extremities is quite characteristic, it is not sufficient in itself to establish a diagnosis of tetany, because it occasionally occurs both in hysteria and in organic diseases of the brain.

A positive diagnosis of tetany rests on the proof of abnormal exaggeration of the mechanical and galvanic irritability of the nervous system. This overexcitability gives rise to a triad of symptoms, which are present either singly or together in the intervals of freedom, and are therefore distinguished by the term "*latent*" tetany from "*manifest*" tetany, the latter being characterized by the spontaneous attacks of convulsions in the extremities (carpopedal convulsions).

Fig. 61.



Manifest tetany of all the extremities. The upper extremities are in the "begging position." Girl four years of age.

The latent symptoms of tetany are: Trousseau's, Chvosteck's, and Erb's phenomena.

Trousseau's phenomenon consists in the fact that pressure on the nerve trunks in the internal bicipital groove, or by elastic constriction of the arms brings on an attack. The constriction, which must be great enough to produce cyanosis of the distal portion of the extremity, must be kept up for from one to several minutes before a convulsion makes its appearance, and the procedure is attended with some pain. The phenomenon, however, is the most fugacious of the three named, and while its presence is a proof of tetany, its absence is of no significance.

Chvosteck's, or the *facial phenomenon* (erroneously called facial reflex) is, like the preceding, an expression of increased mechanical irritability of the nerves. When the facial nerve is tapped at a certain point on the cheek, about midway between the zygomatic process and the angle of the mouth, lightning-like contractions are produced in the entire region supplied by the branches of the facial nerve which is affected by the blow.

If the increase in irritability is very great, active contractions can be elicited by merely stroking the cheek (Schultze's phenomenon) instead of tapping it lightly with the percussion hammer. On the other hand, if the increase in irritability is slight, the contractions are feeble and often appear only in one branch of the facial. It is obvious that the phenomenon can be elicited with difficulty, or not at all, if the child is crying.*

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* Escherich's mouth phenomenon and Thiemich's lip phenomenon, both of which consist in the main of a protrusion of the mouth in the form of a snout on tapping the orbicularis oris, have nothing to do with tetany. They probably bear a close relation to the sucking reflex in the healthy young infant, and in older children that are somnolent or have been injured by some cerebral disease.

The pathognomonic importance of the facial phenomenon is much greater, in our opinion, than is usually believed, a point which will be discussed again later.

The question of determining the mechanical overexcitability in other peripheral nerves has already been discussed.

Erb's phenomenon consists in overexcitability of the peripheral nervous system to a galvanic current, and in formulating the characteristic contraction law of Thiemich and Mann is the most constant and most sensitive indicator for determining the abnormal irritability which is the basal condition, as we have already explained above. It should be emphasized, however, that the galvanic overexcitability need not necessarily be of exactly the same degree in all the nerves at the moment of examination, and that its intensity—in the median nerve, for instance—is not always proportional to the clinical manifestations and the remaining latent symptoms.

The subdivisions of the latent symptoms into obligate (Thiemich's and Erb's phenomenon) and facultative (mechanical excitability, facial phenomenon or laryngospasm) which is practiced by many authorities, lacks sufficient justification and ought to be discarded; but as the expression spasmophile diathesis is more descriptive and more comprehensive, it would be advisable in the future to restrict the term latent tetany or tetanoid conditions to those cases which exhibit the Trousseau phenomenon.

The **clinical course** of tetany exhibits many variations. As a rule, the individual attack does not last more than a few hours, although it may continue for 12 hours or an entire day; the convulsion then usually relaxes, but is repeated after an interval of a few hours, and so on for several days, with several intervals of freedom. The entire duration rarely exceeds two or three days; at least, we observed such a clinical course even at a time when the remedies at our command in all probability did not influence the course of the disease as markedly as we have now learned to do.

While the above course is observed in the majority of the cases there is a small minority in which the carpopedal convulsion loses more or less of its intermittent character and produces a permanent spasm. In these cases the demonstration of latent symptoms, particularly the most delicate of these symptoms, namely, overexcitability, is of decisive importance in deciding the question whether phenomena of this kind may be regarded as tetany or not. It is almost needless to say after what has been stated that, with Fleiner and many others, we hold fast to the view that convulsions in which over-electrical excitability is absent have nothing to do with tetany. According to this view we include under the head of tetany, first, those rare cases characterized by great severity which others as well as myself have observed beyond any question of doubt, and in which the individual attacks lasted many days, while the entire duration of the disease was measured by months.

Second, the cases of general *hypertonia* which occur after certain nutritional disturbances (particularly the condition produced by artificial feeding) and which Gregor first studied and described with regard to their electrical behavior. On the other hand, most cases of general muscular hypertrophy in sick infants, and the conditions described by Escherich as pseudotetanus have nothing whatever to do with tetany.

For practical reasons, we shall here append a description of these conditions, although they exhibit only a superficial and momentary resemblance to tetany.

General Muscular Hypertonias Without Spasmophilia.—The muscular hypertionas—also known as persistent spasm (Zappert) or "myotonia of the newborn" (Hochsinger*)—have been recognized since the appearance of Czerny and Moser's articles on the subject as a frequent symptom of severe nutritional disturbances. The convulsions which may predominate either in the flexor or in the extensor muscles vary in intensity and in duration from a few days to several weeks, but are *never intermittent.* The position of the arms and legs may simulate that of tetany; indeed pressure on the nerves and vessels in the bicipital groove may cause an increase in the muscular tone. At the same time, the child usually makes a fist, a phenomenon to which Hochsinger attributes undue importance. He speaks of a fist phenomenon which he likens to the Trousseau phenomenon. But, although Hochsinger contends that the two symptoms are frequently confused, there is no reason to think that the confusion has ever led to an error in diagnosis. These prominent spasms practically always occur in children only a few weeks or months old; very rarely during the period of childhood which furnishes the chief contingent of tetanic patients (compare page 316); the convulsions almost without exception occur in children suffering from acute septic processes and exhibiting other symptoms of cerebral irritation or palsy. Nothing positive is known in regard to the pathogenesis of these conditions. The treatment is the same as that of the primary disease.

This condition must be differentiated from certain similar conditions which occur in infants suffering from chronic nutritional disturbances without gastroenteric phenomena in the intermediary metabolism. In such infants similar permanent spasms occur not infrequently in association with galvanic overexcitability; they disappear gradually when the infant is placed on breast-milk and return whenever an attempt is made to return to artificial feeding. This observation, which we owe to Gregors, is of fundamental importance and will be referred to again.

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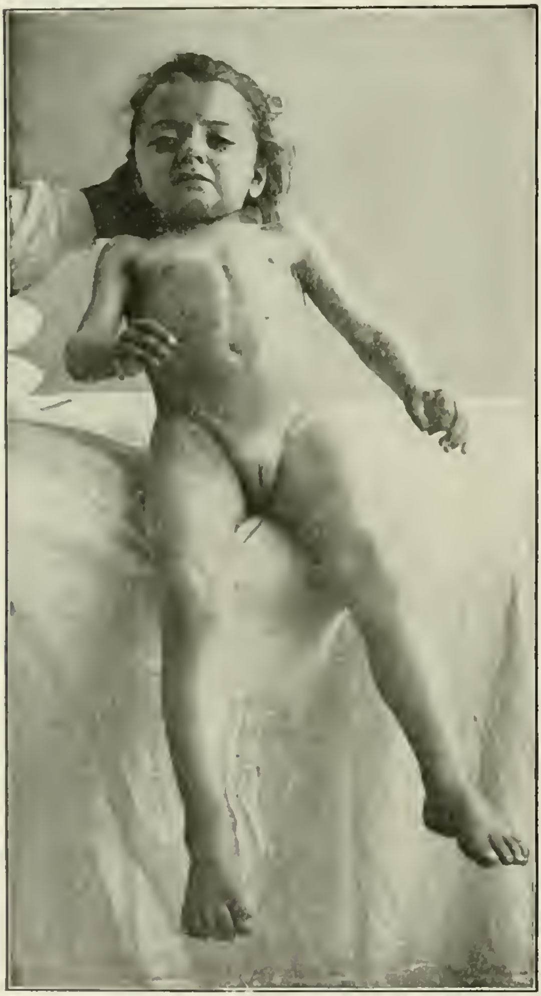
* The selection of this term is unfortunate and Hochsinger in his work on "Krampfe der Kinder" in the "Deutsche Klinik" adds the warning that it is not to be confused with congenital myotonia or Thomsen's disease.

There is another disease which resembles symptomatic hypertonia and permanent spasm and which has been described by Escherich under the term

Pseudotetanus

This condition, which has since been observed by other writers, occurs in children between four and sixteen years of age, preferably boys, and is described by Escherich as follows:—

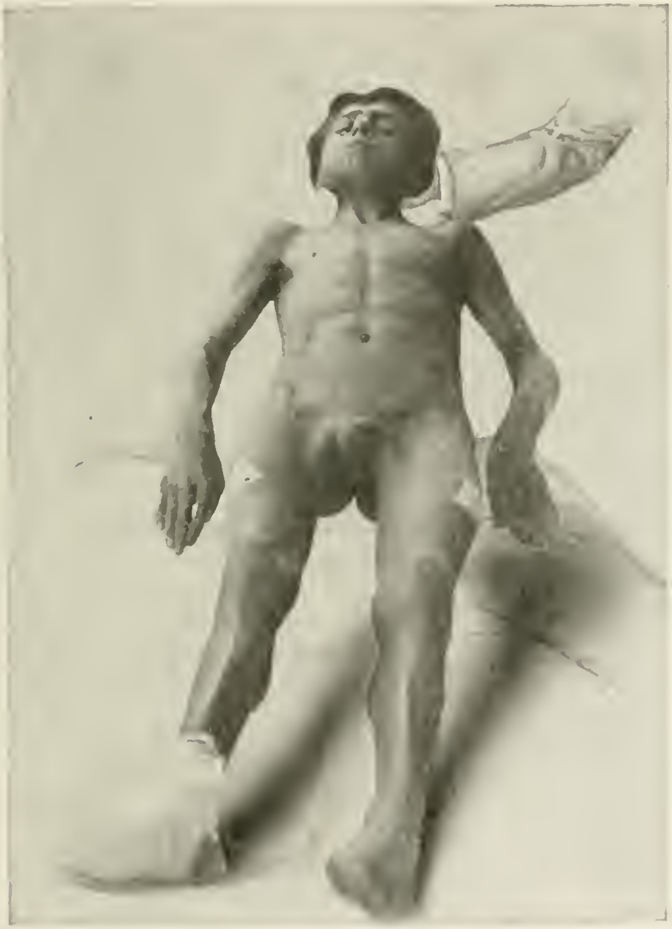
Fig. 62.



Pseudotetanus. Child four and a half years old with pseudotetanus (Escherich). The illustration clearly shows the board-like tension of the lower extremities and the rigidity of the back muscles.

The subject, usually a boy, who has previously been in perfect health and is in no wise tainted by heredity begins to complain of a feeling of stiffness in the legs which interferes with walking so that he has to stay in bed. The rigidity in spite of the rest in bed continues to spread rapidly to the upper portions of the body, the back and the head, and the patient lies in complete extension, immovable and as rigid as a piece of wood. All the muscles of the trunk, the neck and legs are in a state of maximum contraction, they stand out prominently and are as hard as marble. The muscles of the face also are in a state of tonic convulsion producing a peculiar expression of countenance which Soltmann describes as resembling the expression of a person who is blinded by a very bright light. The teeth are tightly clenched and can be separated only a short distance even with the use of considerable force. During rest and when the child is asleep, the rigidity relaxes but never subsides altogether. On the other hand, cooling of the body, noises, touching the patient or psychic exitation produce paroxysms which may lead to still greater muscular contractions, associated sometimes with pain, to opisthotonos, spasm of the diaphragm, dyspnœa, etc. At the height of the disease, paroxysms of this kind may occur spontaneously without any recognizable cause several times a day. The arms and hands and the eyes are not affected and retain full freedom of motion, presenting a marked contrast to the rest of the body which looks as if it were carved out of wood. All the other organs and functions are normal.

Fig. 63.



Pseudotetanus.

This condition begins a few days after the onset of the disease and persists without change for from 3 to 6 weeks when the contractures gradually relax and the patient, after considerable persuasion is at last induced to use his legs again. Complete recovery takes place in from 2 to 4 weeks. Relapses have not been observed.

The nature of this disease has never been fully explained. Escherich called his cases tetany, in spite of the absence of the characteristic overexcitability, but Pfaundler, who recently made an exhaustive study of a new case, rejects Escherich's view for this very reason.

In two of the cases reported in the literature (Kühn, Gomez) the presence of latent symptoms of tetany is expressly mentioned.

Organic diseases of the brain, while they may produce similar pictures, and hysteria can be readily excluded by the general habit of the patient and the fairly typical course, terminating in permanent recovery; but the distinction from genuine traumatic or "rheumatic tetanus" is exceedingly difficult at least during the beginning of the disease. Hence any wound in a child must be carefully searched for the Nicolaier bacillus. A negative finding is of course ambiguous. The absence of fever, which is present as a rule, in pseudotetanus does not exclude infectious tetanus. In his case Pfaundler was unable to find the Nicolaier bacillus, nor could he demonstrate tetanus toxin nor tetanus antitoxin. Nevertheless he considers pseudotetanus an infectious disease closely related in its etiology to traumatic tetanus.

The **treatment** consists in controlling the convulsions and the pain with chloral and bromides or if necessary, injections of morphine. The child must be well nourished, which, owing to the trismus is quite difficult. If necessary, the stomach tube must be employed.

(*b*) LARYNGOSPASM*

The second form of convulsions which at the present day is acknowledged as belonging to the spasmophile diathesis is laryngospasm (*spasmus glottidis*). It is true that this is only a symptom—like the carpopedal cramp in tetany—but it is almost always associated with the phenomenon of spasmophilia, and the cases which form an exception to this rule run a somewhat different course and have a special interpretation. They will be referred to later.

Symptomatology.—Laryngospasm in its milder grades manifests itself merely by a strident, protracted, crowing inspiration. In severe cases, however, the spasmodic closure of the glottis is so complete that symptoms of asphyxia—anxious expression of countenance, cyanosis, short, clonic contractions in the facial muscles and those of the upper extremity, or even, unless the spasm is overcome from time to time, by a forcible inspiration, severe general convulsions may result. Death may occur suddenly in a severe attack of this kind, or the condition, after becoming quite alarming, passes off with a few longdrawn inspirations, and the breathing gradually becomes normal again.

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* In literature the term laryngismus is much used for this affection. We avoid this and restrict the term laryngismus stridulus to the occurrence of audible breathing which occurs in many young infants—congenital laryngeal stridor—which botb from the etiologic and from the clinical standpoint is quite a different disease.

Whereas in a typical laryngospasm, be it mild or severe in degree, there is an unmistakable effort at inspiration, there are cases in which the spasm of the closers of the glottis extends to the diaphragm and the remaining muscles of inspiration. In this way the clinical picture which Kassowitz first described as "expiratory apnœa" is produced. Attacks of this nature place the infant's life in the greatest jeopardy. In favorable cases they end like laryngospasm with a crowing inspiration; or, if the spasm of the glottis has already relaxed, the attack passes off without any sound, and respiration is gradually restored.

According to my experience this expiratory apnœa is the most frequent cause of the cases of sudden death occurring in apparently healthy children, in which the autopsy fails to give sufficient explanation.

The **frequency** of laryngospasmodic attacks is subject to the greatest variation. In one case the attacks may occur singly, while in others they may follow each other in rapid succession. Waking from sleep, weeping, crying, choking, anything in fact that interferes with the normal progress of respiration, and, finally, overfilling of the stomach by a copious meal, favor the occurrence both of laryngospasm and of expiratory apnœa.

The duration of the disease is from a few days to many weeks or months.

The **prognosis**, aside from the possibility of death during the attack, is absolutely favorable.

Relations to Spasmophilia.—When the relations existing between laryngospasm and the spasmophile diathesis were first discovered by Loos and Escherich, the former set up the following axiom: "No laryngospasm without symptoms of tetany." In this form the dictum of Loos was vigorously attacked by the opponents of the theory, and it cannot be denied that it contains a certain measure of exaggeration; but in the great majority of cases, the number of which increases the more often and the more carefully examinations are made, spasm of the glottis is associated with the above-mentioned latent symptoms of the spasmophile condition.

This leaves a small remainder of patients in whom these symptoms cannot be demonstrated in spite of the most careful search. But these cases differ fundamentally by the character of their clinical course from ordinary spasm of the glottis. Some of them belong to the organic diseases of the brain and exhibit, in addition to the laryngospasm, other bulbar and pseudobulbar symptoms (disturbances of deglutition and of the movements of the tongue, constant protrusion of the tongue, and almost regularly profound idiocy); in other cases the laryngospastic attacks ending in apnœa and convulsions are replaced, in the course of the subsequent months or years by typical epileptic attacks; while a very large proportion of the cases which we have had an opportunity to study rested on a foundation of hereditary syphilis. This fact, and the association of well-marked, but non-progressive idiocy, show that the epilepsy must be regarded as symptomatic.

It is worth while to observe that even in rare cases of meningitis there is also a respiratory disturbance similar to laryngospasm, a fact which is explained by the existence of cortical centres for co-ordinated movements of the larynx.

The cases of "holding the breath" without genuine spasm of the glottis, which occur after the age of infancy (so-called paroxysms of rage) never present a single symptom of spasmophilia, but do exhibit the signs of a neuropathic taint (see page 354).

The third variety of convulsions associated etiologically with the spasmophile diathesis is:—

(c) ECLAMPSIA INFANTUM

The **clinical picture** of this condition so closely resembles that of epilepsy that until very recently (Féré) eclampsia was regarded merely as a form of epilepsy characterized by the age of the patient and a favorable clinical course. The attacks consist of a primary tonic, and secondary clonic stage. A kind of aura is also frequently observed. The children become restless, *distrait,* inattentive, and anxious. This stage lasts at most but a few minutes, usually only a few seconds, and is followed by sudden pallor of the face, loss of consciousness, and a tonic convulsion of the muscles of the eye, the face and the extremities. After a few seconds these are replaced by clonic contractions, at first violent, shaking the entire body at each paroxysm as though a powerful electric current were passed through it, to use Soltmann's comparison, with irregular staccato, often audible breathing, approaching a cry, considerable cyanosis and profuse sweat. Gradually the convulsion subsides, and after one or a few minutes the attack terminates in a general relaxation and return to consciousness. If the attack lasts longer than five minutes at the most, the functional character of the convulsions must be called in question.

The similarity to epilepsy is increased by the fact that in many attacks the irritative phenomena are quite overshadowed by the loss of consciousness, and a clinical picture is produced which may quite readily be compared to the petit mal of epileptics.

The **duration** of the individual attack as a rule does not exceed a few minutes, at the expiration of which time the child again appears normal or at most somewhat peevish; the postepileptic stage of somnolence, which is so frequent after epilepsy, is absent.

In severe cases, however, the attacks may be massed and occur at short intervals, so that we are justified in speaking of a status eclampticus analogous to the status epilepticus. The number of attacks that may possibly occur in children the subjects of eclampsia varies within very wide limits. While one patient may not have more than one or a few altogether, another child may go through ten or twenty or even more attacks in the course of a single day, and may never suffer from them again, or may be free for a long time; a third may have attacks at varying intervals for days or weeks. The clinical course depends partly on the severity of the disease and partly on the treatment, which will be discussed later. Death during an eclamptic attack is a comparatively rare event; at least there is no doubt that laryngospasm and expiratory apnœa are much more dangerous in this respect. The usual termination of these attacks is in complete recovery without any permanent damage.

To what extent the mental (psychic) impairment which is sometimes observed in children who have suffered from eclampsia at some previous time is related to the eclamptic attacks, is a difficult question to decide. What we have to say on this point will be found in the chapter on Epilepsy (see page 336).

Eclampsia infantum can be differentiated from other functional or organic convulsions in early childhood by examining the patient during the intervals of freedom, with due consideration of the character and duration of the attacks themselves. Whereas an eclamptic child presents only the symptoms of the spasmophile diathesis during the intervals between the attacks, meningitic, encephalitic or toxic symptoms are found in other conditions. Fever never occurs in uncomplicated eclampsia and is always indicative of the associated infection which may possibly be the primary cause of the convulsions. (See also the chapter on serous meningitis, page 423).

Late Forms.—In the great majority of children eclampsia disappears at the end of the earliest infancy, and the children remain free for a time from manifestations of the spasmophile diathesis, the latent symptoms of which, however, may be demonstrable years later and possibly until adult age. A small minority go through attacks of convulsions usually isolated, but in every respect resembling epilepsy, during the third, fourth, or even the seventh and eighth year of life.

We have not a doubt that the attacks are rarely recognized as "late eclampsia" and differentiated from epilepsy; such a diagnosis is possible only by searching for spasmophile symptoms and taking into account all other accessory circumstances. Since the prognosis of late eclampsia is favorable, in contrast to the unfavorable prognosis of epilepsy, the differentiation of the two conditions is of the greatest importance. These points and the question of the transition between eclampsia and epilepsy will therefore be referred to again in the chapter on Epilepsy.

THE SPASMOPHILE DIATHESIS AS A CLINICAL ENTITY

The three above-described clinical pictures, tetany, laryngospasm, and eclampsia, are quite different in their manifestations, and the only thing that they have in common is the soil on which they grow—the spasmophile diathesis. Another reason, however, for including them in the same group, is that they frequently occur either in combination or in alternation in the same individual, and possess a number of common clinical characteristics in respect to the etiology, the age at which they occur, the peculiar type of children subject to the diseases, etc.

Before taking up these factors in detail, it should be emphasized that in many children the spasmophile diathesis can be demonstrated by repeated careful examination, even though they have never presented symptoms of any of the above-mentioned diseases. The theory first adopted by Thiemich that the presence of galvanic overexcitability is sufficient to characterize a child as pathologic has not yet found general acceptance. On the other hand, the observations which we have made at the Breslau Kinderklinik during a number of years, have tended to strengthen our opinion more and more, and in recent times Finkelstein and H. Neumann have adopted the same view. The explanation which follows accordingly applies to the spasmophile diathesis in general, whether it manifests itself in one form or another, or remains permanently latent.

Galvanic overexcitability, and along with it the remaining symptoms of latency, is so variable and so much affected by the nutrition that a high degree of excitability may be present one day and vanish the next, to be replaced by normal conditions. On the other hand, unless it is influenced by treatment, the spasmophile diathesis may persist for many months with only moderate changes in its intensity. These two facts have a very great theoretical interest, inasmuch as they demonstrate that we are dealing with a functional condition which is not dependent upon the relative degree of development of the peripheral and central nervous system, and is therefore not to be regarded as a phase of development in the sense in which that term is used by Soltmann (see page 297).

In accordance with the above, the pathologic findings that have been described by various authors in tetany and eclampsia cannot be regarded as the material counterpart of the functional disturbance, but merely as secondary and accidental changes.

The observation which, both from the practical, and as we shall see later, from the functional standpoint, is most important for the explanation of the pathogenesis of the spasmophile diathesis, is the fact that it is influenced by the food. The rarity of eclampsia or laryngospasm in breast-fed children, and the fact that both conditions can be cured by feeding the children on breast milk-have long been known to good observers. The exact proof, however, that not only the clinical manifestations but the primary (galvanic) overexcitability is closely dependent on the food, we owe to two series of investigations by Gregor, who discovered two fundamental factors: first, that the withdrawal of food, providing sufficient water is administered (tea diet), causes the disappearance of the most extreme degree of overexcitability in from twenty-four to forty-eight hours; and second, that if the child is afterwards fed on breast-milk, the normal condition continues permanently, whereas artificial feeding is followed by a renewed increase in the galvanic irritability. With this observation as a basis a number of authors proceeded to study the influence of the individual ingredients of artificial food.

Investigations along this line, which are by no means completed, while they have resulted in the postulation of certain definite laws, have also demonstrated the extremely complex nature of the problem. It appears that the influence of diet is by no means a simple, direct influence, but is exerted in some as yet unexplained way through metabolism or the functions of individual organs. This is proved by the fact that the same change of food in children does not always produce the same result, and produces but little effect, or fails altogether, in some cases.

It may be said to be definitely proved that in many cases of spasmophilia there has been more or less severe overfeeding with cow's milk, and the correction of this mistake alone suffices to bring on a gradual restoration to normal conditions. In others the same result is accomplished only by the complete withdrawal of cow's milk. In such cases the child may, if no breast-milk is available, be fed exclusively with oatmeal porridge or a sweetened decoction of flour, and on such a diet the exaggerated galvanic irritability gradually returns to the normal. But the effect is only temporary, and if the farinaceous diet is kept up longer than from one to two weeks at most, the overexcitability may return or the child may even exhibit very extensive nutritional disturbances which are difficult to correct (effects of artificial feeding). The *fats* are comparatively less harmful; while in regard to the various kinds of *sugar* the question still remains undecided.

Another peculiarity that is frequently encountered in children with the spasmophile diathesis is *rachitis*. If we adopt the theory which Kassowitz's school quite properly hold with regard to this disease, we shall find rachitic symptoms in the great majority of spasmophile children even though the symptoms may be quite inconspicuous. Kassowitz accordingly regarded the clinical manifestations of spasmophilia as nothing more than the "nervous disturbances of rachitic children," and his example has been followed by others, for example, Escherich, who speaks of "tetany of rachitic children." With regard to the pathogenesis, Kassowitz assumes that the rachitic hyperæmia of the flat cranial bones produces a collateral hyperæmia or circulatory disturbance in the cerebral cortex, which in turn is the basis of the clinical phenomena. Aside from the weakness of his position, it must be said that the theory is directly contradicted by the facts; for, on the one hand, we often see children who are quite free from rachitis and yet suffer from severe manifestations of the spasmophile diathesis; and on the other hand, these phenomena are often permanently absent in many children with severe rachitis, especially of the skull. In fact, such a parallelism between cranial rachitis and the spasmophile phenomena, which logically follows from Kassowitz's theory, certainly does not exist. The frequent coincidence of the two disturbances, which cannot be denied, is probably explicable on the ground that the two are coördinate conditions which develop on the soil of the same chronic metabolic anomalies, and which have many points in common.

The question as to the action of phosphorus will be discussed later.

Kassowitz's theory appeared to receive strong confirmation from the peculiar relationship existing between the frequency of tetany and the different *seasons of the year*. Just as von Jacksch, Frankl-Hochwart and others, found in the case of manifest tetany of adults, so also Loos, Escherich, Ganghofner and others, found that manifest tetany and larynogspasm in children increase during the beginning of winter and gradually reach their highest point in February or March; after which the cases diminish in frequency, until during midsummer the incidence becomes zero. To illustrate, we quote Escherich's statistical table of 240 cases:

Month	I	II	III	IV	V	VI	VII	VIII	IX	X	XI	XII
Number of cases..	29	51	59	45	10	7	0	1	4	2	21	16

Kassowitz pointed out the similar relation to the time of year existing between the incidence of tetany and that of the rachitic affections, and concluded that there must be an intimate connection between the two diseases. When, however, Loos, Ganghofner, Cassel, Fischl and others later investigated this relation, it was found that, although both diseases increase in frequency during the last months of winter and the early spring months, there is no accurate correspondence in that respect. In general the cases of tetany are most frequent in February or March, while the rachitic affections do not attain their maximum until April or May. But this comparatively slight deviation is by no means such a serious objection to Kassowitz's reasoning as the fact that all these statistics in regard to the frequency of rachitis have been declared worthless. I myself agree with the latter view, although I shall not stop to give my reasons, and have therefore omitted a detailed reference to the literature. No one will think of comparing the tetany of adults to rachitis, although statistics may prove that in both conditions the incidence is greatest during the spring.

The fact under consideration itself we believe, on the strength of years of observation, to be so well established that we do not feel even tempted to offer statistics of our own.

In examining the response to galvanic irritation in several hundred children we found that not only the clinical manifestation (tetany, laryngospasm, eclampsia), but a completely latent spasmophile diathesis also was demonstrable much more frequently during the late winter and spring months than in the summer and fall. The same observation was made recently by Finkelstein in a number of hospital cases which he examined very thoroughly. The reason for this relation between the incidence of these diseases and the time of year is not clear.

It appears from the literature on the subject that the *frequency of tetany*, as well as that of laryngospasm, varies greatly in different countries and cities. While in some localities the cases are so frequent at certain times of the year, as to be absolutely "epidemic," they are said to be rare at all times of the year in others. Whether these statements correspond to the facts, or are merely due to errors on the part of the various officials, is beyond our ken; but for the present we feel at liberty to entertain considerable doubt.

Most authors state that spasmophile diseases are much more frequent in the poorer layers of society, and this observation has in fact been utilized as the basis of far-fetched conclusions in regard to the pathogenesis (respiratory diseases, Kassowitz). Personally, I have so often had occasion to observe the manifestations of the spasmophile diathesis in children living under favorable conditions as regards residence and care, that I am inclined to doubt these statements, and by no means regard the tetanoid diseases as characteristic of the proletariat.

Spasmophilia possesses in a high degree the characteristics of *heredity*. Direct inheritance and a family tendency to the disease are extremely characteristic. Of this fact, which was known to Seligmüller, Pott, and others, I have become more and more convinced as my experience has increased, and I believe it is one of the most important and best established data that we have in regard to the disease. From the clinical material of the Breslau Kinderklinik I now have more than a dozen observations relating to families in whom the mother had laryngospasm or eclampsia in her childhood and still shows a well-marked facial phenomenon as a residuary latent symptom; while several of her children presented this or that manifestation of the same disease in their early childhood and on careful examination are found to have latent symptoms. There is no doubt that one or several in a series of children may escape altogether, just as we occasionally encounter in a family of children predisposed to obesity or catarrhal diseases one member that is lean or exhibits no abnormalities whatever; on the other hand, it is much more common to find that three or four children of the same parents (possibly because of the influence of analogous alimentary diseases) develop convulsions or laryngospasm one after the other in the second or third year of life, and eventually succumb to these diseases.

I was once consulted by a woman who was expecting her seventh child, after losing the other six from convulsions. On questioning the woman, who was quite intelligent, I learned that all these children had been normal at birth, and had continued to develop normally until the time of dentition, when they died of epileptic convulsions without the presence of any febrile infectious diseases or any severe gastroenteric symptoms. It is, fortunately, rare to find so many cases of the severest forms in the same family; but a tendency to the milder forms, and those which remain permanently latent and are discovered only by repeated careful examination, can be demonstrated in many families.

It should be added that in a few cases we found that the disease was inherited from the father's side.

The fact that the mothers of eclamptic children quite often present the facial phenomenon attracted the attention of Kassowitz when he went over the polyclinic material, and he utilized this fact in supporting his theory of the respiratory etiology on the ground that the vitiated air in the houses of the poor acts injuriously both on the mother and on the child. But the fact that the same thing is observed under the best hygienic conditions, and among mothers who work in the open air, refutes this assumption.

Whether spasmophile families are to be regarded as neuropathic in the ordinary sense of the term, must remain undecided until more systematic investigations are made. According to my experience, while there is no lack of nervous or other pronouncedly neurasthenic parents or other relatives in these families, they do not by any means constitute the majority. I therefore believe I am justified in stating that spasmophilia is an independent disease and not merely a symptom of a general neuropathic diathesis. Epilepsy in the parents or brothers and sisters of eclamptic patients is absolutely exceptional.

The *age* at which eclampsia and laryngospasm begin is from about the fourth month of life to the end of the second year; in the case of tetany, to the end of the third year. The greatest frequency is observed in the second and third semesters of the infant's life. This applies both to the occurrence of the first manifest phenomena and to their disappearance. There is no doubt whatever that both the occurrence and the disappearance of the symptoms are dependent on the time of the year as well as on the age, as has already been stated. This peculiarity is particularly noticeable in relapses, which are not rare. The course in these cases is usually about as follows: A child, say in January, when it is nine months old, develops eclampsia and goes through a number of attacks during the spring months. During the summer the attacks cease and the child loses its overexcitability. In the following winter, the beginning of the predisposing season, the spasmophilia returns, and quite often the child has one or two convulsive seizures, although the attacks are rarely massed. In a small proportion of children this is again repeated during the third, or even the fourth winter and spring, although the child has by this time attained an age at which the disease is rare and primary attacks are quite exceptional, or have indeed scarcely ever been observed. I have not observed recurrences during the succeeding year in tetany and laryngospasm as frequently as in eclampsia.

In contrast to these cases, which belong almost exclusively to early infancy, we have those cases of eclampsia and manifest tetany, rarely of laryngospasm, which occur in the third to the sixth or eighth year of life and do not represent relapses. In the case of tetany the diagnosis is clear enough; but when eclampsia develops in a healthy child that has never exhibited symptoms of spasmophilia, the recognition of the disease may be much more difficult, and the differential diagnosis, especially of this form of "late eclampsia" from epilepsy, may be of very great importance.

Persistence of the latent diathesis for years or even decades is much more common than recurrence of one of the above manifestations of the disease. The readiest method of determining persistence of the latent diathesis is by examining the patient for the presence of the facial phenomenon, which, according to Thiemich's observations and explanations, even in older children, must not be regarded as a neuropathic stigma, but as a specific latent symptom of tetany or spasmophilia.

The obvious indication to confirm this view by testing the electrical irritability in older children has never been satisfied, because no one has undertaken the laborious task of collecting absolutely correct normal values for later childhood.

ETIOLOGY AND PATHOGENESIS OF SPASMOPHILIA

It is necessary to make as sharp a distinction as possible between the causes themselves and the conditions which favor the development of the diathesis on the one hand, and the clinical phenomena to which the diathesis gives rise on the other.

Many authors seek the cause of tetany and the overexcitability of the nervous system on which it is based in a *general infection*, exactly as has been done in the case of rachitis. The arguments constantly presented are the great frequency of the diseases at certain times of the year, and in certain localities, which practically amounts to an epidemic: but we are without any definite proof, because the facts adduced in support of the argument are either imperfectly established or capable of some other interpretation. In the absence of complications, the disease runs an absolutely afebrile course, so that there is nothing which could suggest the idea of infectious disease to an unprejudiced observer.

Kassowitz's theory of a *respiratory injury*, the weak points of which we have already pointed out, is also based on the massing of the cases which is observed both as to time and place; nor is the theory of a status lymphaticus, which was first promulgated by Paltauf and by Escherich, applied to tetany and the tetanoid manifestations altogether satisfactory, for even if we accept the status lymphaticus as a well-defined constitutional anomaly, which is open to grave objection, the condition is found only in a certain proportion of spasmophile children. Many of them in fact are quite lean and imperfectly developed, in sharp contrast to the pasty, "lymphatic" type.

The theory that the functional anomaly of the nervous system, which forms the basis of tetany, is due to a *functional absence of the parathyroid glands or* epithelial corpuscles, rests entirely on theoretical consideration and is devoid of pathologic proof. With regard to this subject, which has recently elicited a great deal of discussion, we shall refer the reader to the comprehensive essays of Biedl and Chvosteck, contenting ourselves with the statement that, while the possibility of producing in animals a convulsive state which rapidly ends in death and has been called tetany by removing all four of the epithelial bodies may be regarded as proven, it has not been demonstrated either that this "tetany" is identical with the tetany of childhood, nor have any anatomical changes in the parathyroid glands been observed in infants the subjects of tetany or spasmophilia.

My own comparatively few histologic examinations of these organs have so far given absolutely negative results.

It is certain from clinical observation that the *quality and quantity of the food* may bring on a spasmophile diathesis, and also cause it to disappear. This is accordingly a factor of indisputable importance, but we are far from understanding its mode of action, and for the present must be contented to attribute spasmophilia to some unknown metabolic disturbance.

Experimental physiology has taught us that the irritability of a peripheral nerve can be influenced by salt solutions capable of modifying osmosis, and this suggests the possibility that we may be dealing with a *disturbance of salt metabolism*. With this in mind Czerny instituted a series of chemical examinations of brains, which was carried out by Quest, and which showed a diminution in the calcium salts in the brains of tetanic children. The study of the calcium salts was suggested by Sabbatani's discovery that the irritability of the cerebral cortex to the electric current increases as the calcium content diminishes. If subsequent investigations on a larger scale—which in view of the fact that the gray and the white substances do not share equally and constantly in the building up of the growing brain seem most desirable—should show that Quest's findings have the importance of a law, this would give us another clue to the explanation of spasmophilia and its dependence on diet.

Finkelstein assumes that "the abnormal changes that take place in the bodies which are normally produced in the catabolism of the food and in a healthy child are at once rendered harmless, are the source of the symptoms." It is probable that the disturbance has nothing to do with casein, milk-fat, and sugar, since no harm appears to be done by adding these substances (casein in the form of plasmon or nutrose) to a diet consisting of substances (breast-milk, flour, etc.) which tend to diminish irritability. Undoubtedly, however, whey has the same effect as cow's milk in increasing the irritability, and it may therefore be inferred that it contains in solution a substance which is in some way concerned in the production of the anomaly.

Finkelstein's contention that cow's milk is almost invariably harmful is not quite in accord with my own experience, for I have seen symptoms of spasmophilia in breast-fed children who received either nothing but breast-milk, or, in addition to breast-milk, nourishment entirely free from cow's milk (as, for example, rolls soaked in water, with butter and sugar, or soup made with flour). However, cases of this kind are rare exceptions, and we are not inclined on their account to deny the importance of diet.

Finkelstein correctly pointed out that the "army of children afflicted with the spasmophile diathesis can be divided into two different types, which are closely connected by intermediate forms." One type is seen in the obese, overfed child, in whom, as a rule, a cure can readily be effected by cutting down the diet; the other finds its representative in the lean, the subjects of chronic gastro-enteric disease, in whom spasmophilia cannot be prevented by restricting the amount of food and is not always curable.

This very knowledge, that the spasmophile affections vary in their manifestations, their degree of severity and clinical course, should prevent us from neglecting other factors of etiologic importance which we learn by clinical observation and which we are in danger of overlooking because of the importance we attach to the influence of diet.

One of these factors, direct homologous transmission of spasmophilia from parents to children, has already been mentioned. The significance of this factor is beautifully illustrated by the interesting observation of Finkelstein that children who, without any demonstrable alimentary weakness or, at the most, very slight symptoms of indigestion, acquire spasmophilia, are born of mothers who have themselves had spasmophilia in their childhood and in some instances still present distinct latent symptoms.

Finally, it is probable that the spasmophile diathesis may be produced by protracted diseases of the respiratory organs leading to cachexia, suppurative processes and the like.

Having thus attempted to make clear the predisposing causes of the spasmophile diathesis according to our present knowledge, let us inquire how and in what manner, in a given case of spasmophilia, the individual clinical manifestations (tetany, eclampsia, laryngospasm, expiratory apnœa) are produced. The obvious answer is by reflex action. It will be remembered that Soltmann used the terms "spasmophilia" and "increased predisposition to reflex irritation" synonymously; but it is difficult to make this interpretation harmonize with the absence of exaggeration of the tendon, cutaneous, and mucous membrane reflexes, which ought to be present regularly or at least frequently. Clinical observation supports Thiemich's theory that, in the case of laryngospasm at least, and probably also of eclampsia, some disturbance of the respiration and therefore of the normal ventilation of the blood (oxygenation) is the most important etiologic factor. An attack may be brought on by crying from any cause, by the reflex cessation of respiration which accompanies depression of the tongue to allow inspection of the pharynx, or the introduction of a stomach tube, the practical importance of which requires no further elucidation. It is possible that *overfilling* of the stomach by a too copious meal, which clinical observation would naturally incline us to regard as a cause of convulsions, may act in a similar manner. The cases of sudden death from arrest of the heart in spasmophile children occur so very frequently after a copious meal that we cannot regard it merely as a coincidence.

Reflex irritation in any part of the body cannot be regarded as the immediate cause of the convulsions. It would be almost superfluous to state in so many words that we do not acknowledge eruption of a tooth as a possible cause of the convulsions, were it not that quite recently some authors (Spiegelberg and Bendix for instance) have again taken up this fallacy, which had been successfully vanquished by Fleischmann, Kassowitz and others.

Finally, *fever* must be mentioned as a possible exciting cause. The significance of this factor is very difficult to estimate correctly, although clinical observation shows that fever is frequently coincident with the occurrence of the convulsions. The time-honored theory that convulsions take the place of chills in infants is untenable, for we note that the so-called fever convulsions occur practically without exception in children of a spasmophile diathesis, and furthermore also in mild infections, such as varicella, and in association with slight febrile elevations.

The **diagnosis** and **prognosis** have been sufficiently discussed in the preceding, and all that remains is a short discussion of the

TREATMENT OF SPASMOPHILE DISEASES

As a rule, the acute convulsive paroxysm requires no treatment because it usually terminates before any remedy can be applied. If there is high fever, a tepid bath may be useful; otherwise baths of every kind are useless and only disturb the rest which is so imperative. A narcotic is indicated only when the convulsions are protracted or recur in great numbers, separated by short remissions. For this purpose, we recommend chloral hydrate, at least 0.5 gram (8 minims) of a 2 per cent. solution at a dose, per rectum, the injection being allowed to remain for some time. The drug always acts in from five to ten minutes, and the effect usually lasts from six to eight hours. We have had no personal experience with inhalations of chloroform, which have been recommended by Henoch and others. When the breathing threatens to stop, artificial respiration must be instituted as soon as the muscles relax.

Immediately after the attack is over, measures must be adopted to combat the excessive overexcitability of the nervous system. Among these may be mentioned: Evacuation of the bowels with a purgative (I prefer castor oil, two teaspoonfuls at a dose, to calomel, which is so generally popular), and a tea diet; or, in older infants, oatmeal porridge without any milk, for one of two days. On this diet the spasmophilia usually disappears. When this has been accomplished breast-feeding should, if possible, be instituted at once (at least in younger infants). If this is not feasible, a farinaceous diet offers the best protection against a return of the overexcitability; but owing to the danger of injurious effects from farinaceous feeding (Czerny) this regime cannot be kept up longer than about a week, and must then be cautiously replaced by a milk diet. In a general way it is advisable to cut down the food to a minimum for some time, and rather to forego an increase of weight for several weeks than run the risk of a relapse.

In older infants the restricted diet may be supplemented by milks and soups, and finely divided soft vegetables.

While the overexcitability, after it has once been removed by initial evacuations of the gastro-enteric tract, does not as a rule return if the child is fed on breast-milk, there is no form of artificial feeding that will permanently guard the child against overexcitability. It is possible, however, to keep the overexcitability of the nervous system within bounds and to prevent the occurrence of further manifest symptoms.

The prophylactic importance of this observation is obvious.

In the case of ill-nourished children with chronic gastro-enteric disease the dietetic treatment of spasmophilia presents considerable difficulty. In such cases, which are fortunately comparatively rare, the most important indication is to improve the general condition, ignoring the spasmophilia altogether. Such a policy is quite justified since in these children spasmophilia very rarely brings on any severe accidents that would threaten the infant's life.

In every case the *medicinal* treatment is of very little importance compared to the dietetic management, since the continuous administration of narcotics is hardly justifiable.

A few words must be devoted to phosphorus, which was first recommended by Kassowitz, both for the treatment of rachitic bone changes and for the rapid removal of the "nervous complications" of rachitis, and since his time has been extensively used and recommended by other observers. The adherents (Finkelstein, for example) of phosphorus themselves acknowledge that the drug is effective only in the form of phosphorus-codliver oil which is customarily given. In any other form it is useless, and it must therefore always be prescribed as phosphorus-codliver oil.

How much of the effect is to be ascribed to the phosphorus and how much to the codliver oil is difficult to determine. Finkelstein found that codliver oil alone exerted but a "questionable influence, and that only in exceptional cases." Personally, however, I am inclined to think that codliver oil without phosphorus is more efficacious than Finkelstein believes. With regard to the combination of raw milk and codliver oil, recently recommended by Finkelstein, I have not as yet had sufficient experience to express an opinion. In one severe case of eclampsia and laryngospasm the combination failed to have any effect.

For the present we believe that, aside from regulating the diet, the best thing that can be done is to order phosphorus-codliver oil, which also enjoys a good reputation among the general public.

NODDING SPASM*

The most conspicuous **symptom** of this disease is a more or less continuous rotating, nodding or shaking of the head. The combination of these movements of the head with nystagmus which, while it is not observed in every stage of the disease, is found practically always if the case remains under observation for any length of time—as well as with a number of other peculiarities that return again and again in patients of this type and which we shall describe later, will justify the description of nodding spasm as a separate clinical picture. The differentiation from other diseases which resemble it superficially, but possess an entirely different pathogenesis as well as prognosis, we owe in the main to the work of Raudnitz, to which later contributions by Ausch, Thomson and others, have added but little. Raudnitz, who made a careful analysis of the clinical picture describes it in the following words:—

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*Synonyms: Head-shaking. Head-jerking. Head-nodding. Head-rotating. Gyrospasm. Spasmus nutans, etc.

"The children are at most three years of age, generally between the sixth and twelfth month of life. At this time the movements of the head, consisting of nodding, shaking or rotation, make their appearance; the excursions are quite limited, and the velocity is barely equal to that of the pendulum of a clock. The trunk takes part in the movements only to the extent of responding by a slight, purely mechanical movement to the nodding of the head. Movements similar to those of the head do not occur in the face (except about the eyes), in the trunk or in the extremities. An oblique position and lagging of the head when the eyes are turned in certain directions, are observed in some cases. At the height of the disease there is always nystagmus, which very frequently is confined to or at least more pronounced in one eye. Nystagmus appears later than the other phenomena and, as a rule, disappears earlier than the convulsive movement of the head; but there are cases in which nystagmus is the most prominent feature, and possibly also some in which it constitutes the only symptom. Nystagmus and movements of the head very frequently alternate, the former occurring during intentional or enforced rest of the head.

Peculiar temporary positions of the eyes, adduction, and more rarely abduction of one eye, and convulsive movements of the lids are of frequent occurrence; lachrymation is rare. All these phenomena, which disappear during sleep, bear a distinct relation to the movements of the eyes, quite frequently to movements in certain directions only. The convulsive movements and oblique position of the head cease if, in cases in which the nystagmus is confined to, or most prominent in one eye, the affected eye is bandaged or, in cases with bilateral nystagmus, if both eyes are bandaged. Movements of the head coming on when the eyes are bandaged are absolutely normal. The phenomena of nodding spasms are not attended by any disturbance of consciousness, nor are they followed by exhaustion. Other nervous symptoms are absent in the majority of cases, and the mental development does not appear to be injuriously affected. The disease ends in complete recovery, but may continue for two years, during which time there are distinct relapses or exacerbations."

When we compare Raudnitz's masterly description of the disease and its course with former descriptions, we note that the chief progress lies in the recognition of the fact that the movements of the head and of the eyes depend on the child's desire to look in certain directions.

Raudnitz's observations impressed him with the importance of the etiologic influence of dark lodgings and insufficient or one-sided illumination in the production of the disease. In the majority of these cases he found that the children were forced, by the position which they habitually occupied and by the insufficient illumination—in order to turn toward the window, or whatever other scanty source of light there was, or to examine a plaything which they held in their hands—to turn the eyes constantly to one side, or to rotate the eyeballs upward. This, in his opinion, is the manner in which the motor disturbance is brought about, and it is analogous to the nystagmus of miners. In support of this theory Raudnitz cites the observation, which has also been made by others, that the disease usually begins during the dark months of winter.

Since the disease, however, appears only in a very small percentage of all children who are surrounded by the unfavorable conditions mentioned, we must obviously assume an individual predisposition, in regard to the exact nature of which we can at present do no more than speculate. In some cases it is found that the children have been gradually reduced by some chronic nutritional disturbance or febrile disease. Most of these are more or less rachitic; but it is not possible to demonstrate any neuropathic taint, nor does the condition appear to lead to any other nervous disease. Raudnitz offers the ingenious suggestion that certain dynamic conditions in the eye muscles are the chief cause. If the muscles are naturally abnormal, even such a slight injury as the lack of light might be enough to produce the nodding spasm and nystagmus.

The **treatment** of nodding spasm consists in securing better illumination and in improving the general condition of the children, if it is unfavorable, by suitable feeding. If this can be done, the **prognosis** is always absolutely favorable.

The only condition that might present diagnostic difficulties is juvenile or congenital nystagmus if, as occasionally happens, it is associated with isolated movements and oblique position of the head. The differential diagnosis can usually be made at once, since in juvenile nystagmus there is usually some definite cause (central macula, cataract, amblyopia, and the like). At any rate, the subsequent course of the disease will serve to clear up any doubt.

Similar tremors or nodding movements of the head, which may occur at the beginning of some organic nervous disease such as tuberculous meningitis (Demme), are at no time accompanied by nystagmus and, if due attention is paid to the concomitant symptoms, should not be difficult to diagnose. The same is true of multiple, insular sclerosis and Freidreich's ataxia, in which, besides, the patients are always older.

Certain swaying or rolling movements observed in psychically abnormal and imbecile children sometimes exhibit great similarity to nodding spasm; but nystagmus is absent, and the movements themselves are less constant and not so much confined to the head. Moreover, the feeble-mindedness reveals itself early by the objective signs. These movements will be described at greater length in the section on Sterotypias.

Finally, it should be briefly mentioned that the conditions described as nodding epilepsy, or beckoning, saluting or salaam spasms must be sharply distinguished from nodding spasm, to which they also present some superficial resemblance. These anomalies belong to epilepsy.

CHOREA MINOR

The prominent symptom in the disease which we designate chorea minor, infectious or Sydenham's chorea is the choreic motor disturbance. As recently analyzed by Förster, it is compounded of peculiar reaching "spontaneous movements" and "choreic disturbances of coördination." In this way the well-known characteristic picture is produced.

The character of the movements does not, however, in itself suffice for a diagnosis of minor chorea; the choreic motor disturbance is merely a symptom which also occurs in other organic functional and nervous diseases.

Brief mention may be made of simple chronic and progressive chronic family (Huntington's) chorea; hysterical chorea; and the *chorée variable des dégénerés* (Brissaud), which develops in degenerates; electrical chorea; posthemiplegic chorea; and similar more or less partial forms of chorea. They will be referred to again at greater length in discussing the differential diagnosis.

Clinical Picture.—Minor chorea is a subacute disease of childhood and early youth, which ends in recovery after a few weeks or months. The choreic motor disturbance, as a rule, affects all the voluntary muscles, although frequently it is more prominent on one side of the body than on the other; the disturbance is never confined to a single extremity; it is never associated with spastic cerebral palsies, like postapoplectic chorea; on the contrary, the cases, without exception, exhibit a flaccid, partial paralysis (chorea mollis or paralytica). During sleep the choreic movements cease. The disposition of the patients is usually affected; they are given to sudden changes of mood, are readily moved to laughter or tears; there is an utter want of concentration and inability to fix the attention or perform any mental work; the children are excitable and easily frightened. Psychic conditions are not rare in individuals who have passed the age of childhood.

A definite relation has also been established between chorea minor, rheumatism and endocarditis; this connection is not observed in other forms of chorea and strongly suggests that chorea is to be regarded as an infectious disease.

Our reason for adhering to the general custom of describing the disease among the functional nervous diseases is that the symptoms of bacterial or microbic infection, such as fever epidemicity, etc., are absent or very insignificant, so that it is apparently at least a pure neurosis.

The **pathologic findings** in the nervous system that have been reported so far, aside from the cardiac lesions, which must for the present be disregarded, are not of the character or importance to alter our opinion in regard to the nature of the disease. They are either so vague as to lack any pathologic dignity, besides being inconstant, as, for example, the so-called chorea corpuscles; or they are merely the results of the complicating septic or endocarditic diseases, such, for example, as the emboli in the small arteries of the brain, and cannot therefore be regarded as the anatomical counterpart of the clinical symptoms in uncomplicated cases.

In spite of the absence of well-defined pathologic changes in Sydenham's chorea, the fact that the motor disturbances coincide completely with those which exist in symptomatic chorea permits us to draw at least a cautious conclusion in regard to the **anatomical localization** of the unknown pathologic changes. Bonhöffer was the first to observe the combination of choreic spontaneous movements and choreic disturbance of coördination, which are characteristic of chorea minor, in a case of tumor of the crura cerebelli. In agreement with the older statements by Gowers and numerous later investigators, we may therefore assume a toxic infectious lesion of the cerebellum, not sufficient to produce gross and anatomical changes, as the cause of chorea minor. According to our present knowledge the localization of the disease in the cerebellum explains the choreic motor disturbance; but the presence of psychic anomalies, which we are compelled to locate in the cerebrum, proves that the pathologic changes of chorea minor, whatever they may be, are more or less diffuse and involve the entire central nervous system.

It should be added here that Heubner was also impressed by the peculiar character of the choreic motor disturbances and was led to say that "the pathologic irritation to which these movements owe their origin, cannot be referred—or at least exclusively referred—to those regions of the brain which are generally called the motor regions, *i.e.*, the anterior central convolution and the pyramidal tract."

Mode of Onset.—The motor disturbance in chorea usually begins gradually. Spontaneous movements first make their appearance, and disturb the child's muscles, either when they are at rest or during a purposive movement.

The child is unable to sit still, wriggles, makes faces, drums on the table with its fingers, picks at its clothes, shuffles its feet, drops things on the floor and breaks them, etc. These involuntary movements, as well as the movements which accompany every intentional movement that requires the slightest exertion, render the patient ridiculous, an object of general derision. Quite frequently the children are not only teased, but punished for misbehavior, until the true state of the case is finally recognized by some intelligent parent or teacher. Sometimes the physician is the first to give the true explanation.

The pathologic nature of the motor disturbances, however, at once becomes clear when the disease is well developed. The child is then in a condition of constant unrest; the play of features is exceedingly lively and variable; and the greatest variety of emotions are expressed in the most exaggerated manner, in rapid succession and without any reference to the particular mood that the child may be in at the time. The tongue and all the muscles that take part in the act of speech are affected by the disturbance, and a characteristic change is observed in the speech. A few syllables or words are pronounced properly; suddenly the voice drops to a barely perceptible whisper or ceases altogether while the child is trying to pronounce several words, and in its vain attempts to speak it indulges in a number of more or less droll, fantastic or even alarming gestures. When the obstacle has been overcome, a few words are again pronounced correctly, or nearly correctly, and so the comedy goes on until the child bursts into tears and gives up the attempt at conversation, the futility of which has a most depressing influence.

The breathing is affected in the same way, and often becomes sighing and interrupted.

The child is unable to perform the smallest movements with the extremities, for the motor impulse gets off the track and reaches muscles which it was never intended to set in motion. Thus, for example, instead of opposing the thumb as the child may have intended to do, it may flex the hand or spread out the fingers, or perform some other similar movement until suddenly the desired movement is performed, apparently by accident. When, after laborious attempts, the child has finally succeeded in assuming the desired position, it is unable to maintain it more than a few seconds, and has to begin all over again.

At first the child still retains the power of sitting, walking, standing, etc.; but at the height of the disease these static functions are impossible on account of the instability of the movements. The gait is uncertain, and the child not only stumbles but even falls down when it attempts to stand or take a few steps, unless it is supported. Even in bed the movements which accompany every spontaneous intentional muscular action become so marked in severe cases that the patients fling themselves about on the bed in a state of what the French call "folie musculaire," and often sustain abrasions or even serious contusions, or fall out of bed.

Objective examination in pronounced cases of chorea usually shows a very marked diminution of the muscle tone (Bonhöffer). Thus, during the attempt to raise the child by the shoulders the shoulder girdle goes up to the level of the ears. In the extremities also the uncontrolled swaying movements induced by passive walking are quite conspicuous. Normally there is no diminution of gross motor strength.

Paresis, whether mild or severe, is quite rare in choreic patients. Before the appearance of the choreic movements paresis may develop gradually in the form of monoplegia or hemiplegia and may for a long time dominate the clinical picture (limp chorea or chorée molle); or they may develop in the extremity or side of the body which had been the seat of choreic movements and replace these movements; and as they disappear the chorea may again assert itself (paralysie de la chorée). There is no sharp division between the two forms (Rindfleisch). The prognosis of paretic chorea is always favorable.

The **development of the disease** is generally subacute, occupying from one to several weeks. Fortunately the severe forms which we have just described are by no means frequent. In most of the cases the disease does not progress beyond a mild grade of muscular unrest, and then remains stationary for weeks or even months.

When the **course** is afebrile, the general health of the child is often remarkably little affected, particularly if the child is able to take sufficient food, sleeps soundly at night, and sleep is not disturbed by muscular unrest. In children who are naturally delicate or markedly neuropathic, anorexia often develops early, along with the psychic depression, and may render the condition alarming. Severe cases are complicated by insomnia because the movements prevent the child from falling into a deep sleep.

The **duration** of the entire disease, up to the complete disappearance of muscular unrest, may be said to be from one and a half to two months as a minimum, and from six to eight months as a maximum. A longer duration, say from one to two years, while possible, should awaken a suspicion that the illness is not a true chorea minor, but a symptomatic form.

Relapses are quite frequent in chorea. One or two may be observed in the same individual, and it appears to make no difference whether the first attack is mild or severe. As a rule relapses are milder and of shorter duration than the primary attack, but with each relapse there is increasing danger of the development of an endocarditis. It is to be noted, however, that some at least of the cases of apparently secondary attacks are not genuine chorea, and represent hysterical autoimitation. Even the most careful study of the clinical picture may not enable the observer to distinguish between the two conditions, a positive decision being arrived at only by noting the prompt effect of suggestive treatment.

The relations of chorea to rheumatism and diseases of the heart are exceedingly important. Since the middle of the nineteenth century this subject has aroused the interest of numerous investigators, but their labors have failed to bring about any unity of opinion or to clear up the question entirely. It may be stated that it is now generally recognized that in a large percentage of cases of chorea the heart becomes involved in the course of the disease. The cardiac complication may be confined to the presence of a slight blowing or breathing systolic murmur, heard specially at the apex; quite frequently, however, the accentuation of the second pulmonary sound and the persistence of the cardiac murmur after the chorea has run its course point to the existence of organic endocarditic disease. In harmony with these findings in the moderately severe cases, we find almost without exception in the autopsies of severe cases small exerescences of a granular nature, barely large enough to be visible and affecting especially the mitral valve, which prove that there was organic disease of the endocardium. The vegetations are so small, however, that they produce no disturbances in the mechanism of the circulation and as a rule do not give rise to auscultatory phenomena, or at the most to very slight physical signs in the heart. When we remember also that occasionally the endocardium is found at the autopsy to be perfectly normal even in cases in which there was a heart murmur during life, we shall be forced, with Wollenberg, to adopt the stand that statistics in regard to the frequency of heart murmurs and cardiac lesions in chorea are of very little value, unless the patients are followed up, as has been done by Osler, Heinrich Meyer and others. When this is done, it is found that about half of all choreic patients ultimately present positive clinical signs of chronic valvular disease.

In view of this close relation between chorea and organic disease of the heart, which is by no means clear to our understanding, the relation existing between chorea and rheumatism has for a long time been made the subject of investigation. Since we know that chorea chiefly occurs during childhood and rheumatism preferably affects individuals who have passed the age of puberty, it seems desirable to investigate, on the one hand, how many choreic patients present symptoms of rheumatism either before or during their attack of chorea; and, on the other hand, to determine how many choreic patients are attacked by rheumatism when they attain adolescence. When this is done, it is found that the percentage of these cases is also very high, much too high to be explained by mere coincidence. In view of this fact one is forced to adopt the view which H. Meyer, Heubner and others have taken, that the vague "rheumatic" joint pains, accompanied by depression and a general feeling of malaise, which not infrequently make their appearance before or during the choreic attack must be regarded as manifestations of the rheumatic infection.

It is obvious that the course of chorea is very much influenced by the complicating rheumatism, or endo- or pericarditis, or other rheumatic disease, such as pleurisy, which, as we know, are quite frequent. They furnish an explanation for the fact that chorea, in spite of its generally favorable prognosis, may, like any other infectious disease, occasionally present septic phenomena and a fatal termination.

Death from chorea is a rare event and occurs only in from two to three per cent. of the cases. In some of these death is directly due to the cardiac disease and its consequences; other fatal cases, however, are so-called pure uncomplicated cases of chorea. The latter chiefly deserve attention, although they are rarer in childhood than at a later period, up to the age of about twenty years. Only one or two fatal cases under the age of seven years have been reported (Richon), and practically none of boys at any age.

The danger of sudden death, *i. e.*, within a few hours or days, appears to be greater in those patients in whom the psychic symptoms, such as great irritability, violent temper, etc., are most prominent, a phenomenon which recalls the well-known fact that in nervous individuals the danger of sudden and unexpected death must always be reckoned with, even when they are suffering only from a comparatively mild organic disease.

Unfavorable symptoms in the course of chorea are: Sudden elevation of temperature that cannot be explained by any obvious complication, disturbance of the respiration, a small frequent pulse, pallor, and cyanosis. Such prodromal symptoms, which usually coincide with a great change for the worse in the choreic movements (état de mal choréique, Charcot), are generally followed in a short time by coma and death.

The pathologic findings, aside from the changes in the mitral valve, do not suffice to explain the rapid death and consist in serous meningitis, such as occurs in a great variety of infections and intoxications. It seems justifiable to attribute death to this serous meningitis, particularly as the manner of death bears a close resemblance to that which is observed in "*apoplexia serosa.*"

The *age* at which chorea first makes its appearance varies between 4 and 20 years, the period between the 7th and 13th years of life furnishing the largest number of cases. The reason why chorea is most frequent during this period of life is not easy to find.

Heubner, who regards the choreic movements as mimic movements, attempts to explain the comparative immunity of the first three or four years of life by pointing out that chorea cannot occur in a child that has not yet learned to speak in pantomine in whom, therefore, the corresponding nerve centres are not capable of function and cannot be placed in a state of infection by the toxic or morbid process which is the foundation of chorea.

In regard to *sex*, girls, according to our statistics, appear to be affected more than boys. The proportion is greater in statistics based on clinical material from private practice (one male to 25 females), and smaller when dispensary material is utilized (one male to 1.3–1.6 females). As Wollenberg points out, this indicates that girls are more often attacked by the severe forms, which require hospital treatment, than are boys.

The *time of year* probably has some influence on the frequency of the choreic diseases. It is greater in the cold, wet months, and less in dry, warm weather, a peculiarity which is also common to many diseases that are certainly not infectious and cannot therefore be made the basis of any further conclusions.

Direct homologous heredity [the identical disease] does not play an important part in chorea. Wollenberg states that, according to a number of statistics, heredity is present in about 2 per cent. of the cases, of which about 1.5 per cent. show inheritance from the mother alone. On the other hand, the occurrence of chorea in one member of the family and rheumatism in another is not infrequent.

A general *nervous disposition* is observed somewhat more frequently. Judging from the literature, the percentage of choreic patients with a neuropathic diathesis varies between 20 and 36; but these figures must be considered as representing the lowest limit, since a negative history is obtained in many families that are undoubtedly neurotic. The relations between neuropathia and chorea are by no means clear. If, with Heubner, we regard chorea as a "rheumatic equivalent" as the localization of the rheumatic infection in the nervous system, the question whether rheumatic patients of a nervous type are more apt to develop chorea than those who are not nervous becomes doubly interesting. This question cannot be answered off-hand in the affirmative, at least so far as our experience has gone. On the other hand, it is not to be denied that febrile diseases, or chronic nutritional disturbances which lower the resistance of the entire organism, and therefore of the nervous system also, produce a certain predisposition to the intoxication or infection which manifests itself as chorea.

The question of the influence of nervousness in the production of the disease naturally suggests the etiologic significance of *psychic traumatism*. Although such a traumatism often occurs in the history of choreic children in the form of fright, grief of some kind, or mental or psychic overexertion in school, but little importance is usually attached to these factors in the literature. This may not be altogether justified, since when the disease is well developed, we usually consider complete bodily and psychic rest as the most important factor in the recovery.

From what has been said, it appears that the **diagnosis** of chorea minor is usually easy and sometimes can be made at the first glance. Of the diseases which must be considered in the differential diagnosis, hereditary chorea (chronic progressive, or Huntington's chorea) is excluded at once by the fact that it develops at a later age than childhood. So-called electrical chorea (see page 334) is characterized by the fact that the lightning-like convulsions are confined to certain symmetrical groups of muscles (particularly of the neck, shoulder girdle and arm), while the rest of the muscles escape altogether.

Intra- and extra-uterine organic disease of the brain may, under certain conditions which we do not know, terminate in a condition of muscular unrest closely resembling chorea, instead of the muscular rigidity of the extremities, which is a much more frequent result. Similarly, the movements which accompany every intentional movement in many forms of congenital rigidity of the limbs may exhibit a certain resemblance to choreic movements. The primary disease in both classes of cases is at once revealed by a systematic examination.

The muscular unrest and the twitching, jerking movements and grimaces indulged in by neuropathic children from shyness, and particularly when they know that they are observed, are more apt to be confounded with chorea minor, and this mistake is not uncommon in general medical practice.

A single examination is not always enough to make the distinction absolutely clear, especially if the history is confused and misleading and it is stated that the movements began at a certain definite date, when, as a matter of fact, they were only accidentally noted for the first time by the attendants at that particular time. If one can succeed in diverting the child's attention and fixing it on some other subject, these movements usually disappear, while choreic movements persist. Some assistance is derived from the objective examination, inasmuch as the passive movements in neuropathic children are more apt to be accompanied by heightened muscular tone—they are unable to relax their muscles; while, on the other hand, a pronounced hypotonia is the rule in chorea. The greater the neuropathic condition, the more persistent may the movements become (chorée variable des dégénerés, Brissaud). These cases merge without any sharp lines of division into the large group of maladie des tics. Hysteria may also produce similar clinical pictures.

The **prognosis** of genuine chorea minor has been sufficiently discussed.

The **treatment** of chorea offers a wide field for medical art, but it should be remarked at the outset that as yet we have no specific remedy, and it is probable that our methods of treatment have but little influence on the duration of the disease. On the other hand, we are rather more able to modify the intensity of the irritative symptoms.

In spite of their specific action in articular rheumatism, the salicylates and antipyrin appear to have as little distinct influence on chorea as on the development of cardiac affections, and are accordingly suitable only when chorea is associated with rheumatic or cardiac symptoms. In such a case the drug may be given in doses of 1 gram (15 grains) to a child from five to six years old; and of about 2 grams (30 grains) a day, to one between 10 and 12 years of age (Filatow).

The remedy enjoying by far the greatest popularity is arsenic, in the form of potassium arsenite, Fowler's arsenical solution, or arsenious acid.

As Fowler's solution contains one per cent. of potassium arsenite, the dose of two to five drops, which is usually ordered for a child, three times a day in ascending doses, contains 0.015 Gm. (¼ gr.) of arsenic, which is but little below the official maximum dose of 0.02 (⅓ gr.); nevertheless, quantities considerably in excess of this are given by many authors, for example, Comby, Filatow and others. As arsenious acid in large doses is said to be better borne than Fowler's solution, Comby orders an aqueous solution of arsenious acid, 1:1000, mixing 10 grams, (2½ drams), with six tablespoonfuls of water on the first day, and directing that a tablespoonful be taken every two hours. After each dose the child drinks a little milk; on the second day, 15 grams (3¾ drams); on the third day, 20 grams (5 drams) of the solution are given; and so on until on the seventh day, the daily amount is 40 grams (10 drams) of the solution. After the eighth day the doses are reduced at the same rate, so that in the period of two weeks the child consumes 0.350 grams (5½ grains) of arsenious acid.

If nausea, vomiting or diarrhœa develop, the arsenic is withdrawn for a day or two.

In one of Comby's cases an ascending paralysis, with incontinence of urine and feces, loss of the tendon reflexes and sensation, ending slowly in recovery, occurred four weeks after the arsenic had been stopped; and, as the paralysis could not be ascribed to anything else but the arsenic, Filatow advises that the dose be made only half as large. With Comby, he contends that by this treatment not only the choreic motor disturbances are diminished in intensity, but that the duration of the entire disease is shortened to a few weeks. They admit, however, that this treatment with forced doses of arsenic must be instituted early. The method does not as yet appear to have found any imitators in Germany.

Of other remedies the bromides, chloral and opiates, particularly morphine, may be temporarily used when there is much restlessness.

Baccelli and others recommend camphor monobromate (camphora bromata) 1.0–1.4 Gm. (15–20 gr.) for children about six years old; and 1.2–1.8 Gm. (18–30 gr.) for children about the age of ten.

In the Breslau Children's Hospital we have sometimes seen excellent hypnotic results when there was severe muscular unrest, interfering with sleep, from the use of hyoscine hydrobromate in daily doses of one-half to one milligram, while in other cases the drug failed even when employed in twice as large a dose. At all events, if chloral proves inadequate in severe cases of this kind, scopolamin injections are quite justifiable.

We refrain from mentioning a number of other drugs which have been recommended in the treatment of chorea, because they have not been sufficiently tested.

The most important curative factors in the treatment of chorea, however, are not drugs, but physical methods of treatment. The necessity of absolute rest and avoidance of any excitement has been mentioned. For this reason choreic patients should be confined to their beds, at least as long as there is any pronounced disturbance of the static functions; but to keep them in bed until the complete disappearance of choreic twitching, as most authors recommend, does not seem to me proper in view of the duration of the disease, which may be protracted for months. As yet, however, it is impossible to formulate any positive rules in this respect.

Wet packs of several hours' duration, and warm baths, 32° to 36° C. (89.6° to 94.8° F.) possibly followed by light massage, almost regularly produce an excellent sedative effect. These measures may, if necessary, be repeated several times a day, but the patients must be under careful and constant supervision, even if they are comfortably and securely supported on a sheet in the bath-tub.

It is hardly necessary to state that if the child is very restless it should be protected against injury by padding the sides of the bed, and that due attention must be paid to feeding, cleanliness, the evacuation of the bowels, etc.

Rheumatic and cardiac affections occurring in the course of chorea require the same treatment as under ordinary circumstances; for this, the reader is referred to the corresponding chapters in this book.

During convalescence the child must be kept under careful supervision and not be allowed to take up its work again too soon. Several weeks should elapse before the child is permitted to go back to school.

CHOREA ELECTRICA

This term has been applied to various morbid conditions. The affection described by Dubini does not appear to have been observed outside of Italy and may be omitted, as its characteristics have not been sufficiently defined.

Henoch and Bergeron apply the term *chorea electrica* to a condition characterized chiefly by lightning-like contractions in certain definite muscle groups. Henoch gives the following description of the disease in his text-book:—

"From time to time only twitching movements occur, particularly in the muscles of the back of the neck and of the shoulders (sternocleidomastoid, levatores anguli scapulæ, pectorals and trapezius), but also in other parts of the body, which present the greatest similarity to the contractions produced by an induction current of moderate strength. As a rule, the contractions are feeble and of such short duration that it sometimes requires very careful observation to notice them at all. In all, I have seen at least 30 cases of this kind, which occurred both in boys and in girls, all of them between the ages of nine and fifteen years. When the clothing is removed, the rapid twitching of the individual muscles is distinctly seen and felt in the nude body, and when the tongue is protruded, vermicular movements similar to those seen in ordinary chorea are occasionally observed. Each contraction lasts only an instant, but the intervals are quite variable—from only a few seconds in some cases to several minutes in others, particularly if the patient's attention is distracted. Speech is not disturbed, nor is there any interference with writing, sewing, and the like, unless the act happens to be interrupted by twitching of the arm. One half of the body is some times more markedly attacked than the other. In a girl fifteen years of age, the twitchings were confined to the right half of the body and face, and recurred at such short intervals as to interfere with writing and other work performed with the right hand. The motility was perfectly normal, and other symptoms were absent except for an irregularity of the heart which was occasionally observed. In one case the twitchings persisted during sleep, although with less intensity and at longer intervals. In all the others the choreic movements ceased entirely during sleep, as in ordinary chorea.

In a boy ten years of age, whose entire body was convulsed by these lightning-like twitching movements, while the head escaped almost completely, every convulsive jerk was followed by a convulsive inspiration, indicating involvement of the diaphragm and perhaps also of the glottis; while in a girl, twelve years of age, the twitching movements were frequently accompanied by the utterance of one or two inarticulate sounds (vocal spasm). The child's psychic powers were never impaired any more than the speech, which at the most would be interrupted at the instant of the concussion.

Bergeron makes practically the same statement as Henoch about his cases of chorea electrica, which also occurred in children between the ages of twelve and fourteen. Some of them were anæmic or nervous.

The only difference between Bergeron's cases and those reported by Henoch is that the former rapidly terminated in recovery, while Henoch reports that in his cases treatment was usually without effect. After a critical analysis of all the statements found in the literature, and on the strength of his own experience, Bruns recently suggested that the syndrome described by the term chorea electrica may be divided into three pathogenically different conditions: (1) chorea electrica in the strict sense of the term; (2) hysterical; and (3) a form belonging to epilepsy.

Chorea electrica proper is included by Bruns among the tics which are not pure motor neuroses, since they rest on a foundation of nervous degeneration, and are closely related to hysteria, although not genuine hysteria nor offering the same favorable chances of recovery.

It is in this latter respect that chorea electrica proper, which is to be regarded as a subvariety of tic, differs from the hysterical form. The fact that Bruns' cases rapidly ended in recovery permits us to classify them as hysteria. The therapeutic indications which result from this diagnosis are mentioned in the chapter on Hysteria.

A third variety belongs to epilepsy. The diagnosis is possible only if, along with the isolated lightning-like muscular twitchings, there are or have been typical insults, or such insults develop later. Treatment is then directed against the underlying epilepsy, and in view of the impossibility of differentiating the two first-named varieties from the outset, Bruns quite properly advises that the cases should first be treated as though they were hysterical, *i.e.*, by isolation and "intentional neglect." Hysterical patients almost regularly recover under this treatment, which, on the other hand, does no harm whatever if the patient is a degenerate suffering from a variety of tic. In fact, the diagnosis of hysteria is finally confirmed only by the effects of treatment.

EPILEPSY

Even if we exclude all cases characterized by symptomatic convulsions, which are interpreted as "*acute epilepsy*," and restrict the use of the term "epilepsy" to designate a distinctly chronic disease "manifesting itself either in frequently recurring convulsions with loss of consciousness, or in concomitant symptoms of such attacks, or in psychopathic concomitant or sequential phenomena" (Binswanger), we shall still lack a clear insight into the true nature of these conditions. Especially are we unable in the present state of our knowledge to determine whether we are justified in classifying epilepsy among the functional nervous diseases, or whether we must include it among the organic diseases.

We are also at the present time unable to give a final answer to the question whether so-called primary or genuine epilepsy is identical with the symptomatic or secondary form, as Binswanger believes in contradiction to the old Nothnagel classification. Whereas in genuine epilepsy the somatic or psychic attacks with their concomitant or sequential phenomena for years constituted the entire clinical picture; in symptomatic epilepsy they merely form part of an extensive impairment of the cerebral function (idiocy, infantile cerebral palsies), which depends on organic changes.

It is quite natural to admit, in the case of these large groups of so-called symptomatic epilepsies, that the convulsions are due not to perverted function, but to lesions of the brain that are susceptible of histologic demonstration.

It is not necessary to go into this matter any further, and we refer the reader to Zappert's discussion of the question in the preceding section.

Both forms of epilepsy are observed in childhood, and the necessity or desirability of distinguishing between the two depends on whether there is any marked clinical difference. Although in many cases it is impossible, as Binswanger emphatically points out, to say during life whether one is dealing with an anatomically normal brain, or at least one presenting only a secondary atrophy (genuine epilepsy), or with severe diffuse or focal changes (symptomatic epilepsy), it is nevertheless possible in the great majority of cases to make this distinction, and in our opinion, such a distinction is desirable for many reasons.

In the first place, the *age* at which epilepsy begins, and accordingly the frequency at the various periods of life, is different. A study of the literature reveals in this respect the greatest contradictions, which appear to be difficult to reconcile. Whereas, according to Chaslin, epilepsy most frequently begins between the ages of 7 and 15; or, according to Bouchet and Cazauvielh, between 10 and 15; or 12 and 16 (Beau), H. Neumann finds that epilepsy commences most frequently between the second and fifth year; a little more rarely during the sixth to the tenth; and still more rarely between the tenth and fourteenth year of life. This statement of the statistical discrepancies which, brief as it is, suffices for our purpose, shows that we are dealing with differences that cannot be attributed to chance. It may be suggested that they are due to the difference in the material, the first series of statistics being derived from patients in epileptic or other homes; while Neumann's statistics are based on a children's clinic. This may be admitted, but it also implies a difference as regards diagnosis and prognosis. The cases in which the disease develops early are for the most part cases of secondary epilepsy, and the majority of these patients do not attain the age at which they are sent to institutions because they succumb to the progressive cerebral disease; it is from those who are attacked later in life that the inmates of institutions are mainly drawn.

There is a difference in the clinical type of the first attacks, as well as in the time of their first appearance. In genuine epilepsy the first attack is like the succeeding ones, and is of characteristically short duration. In the secondary epilepsies, the first attack, which often represents the onset of the infectious or toxic cerebral affection, is not infrequently much longer in duration. It may last hours, or even half a day or night. Later in the course of the disease the attacks usually become shorter and more like those of genuine epilepsy. Again, in the symptomatic form the first protracted attack at the beginning of the disease is often replaced by a series of short attacks occurring together during a period of days or weeks; while, on the other hand, in primary epilepsy the occurrence of isolated attacks, separated by more or less regular intervals of weeks or months, is practically the rule.

Secondary epilepsy also reacts differently to the bromide treatment. As a rule, the disease remains practically uninfluenced by the bromides.

After these preliminary remarks, which are made partly for the purpose of orientation and partly to explain the difficulties of the question, we will proceed to give a short description of the **symptomatology.**

The typical attack of *grand mal* is exactly the same in the child as in the adult. In either case the attack may come on unheralded, or may be announced by an aura of variable duration and character. To describe these things, which are found in every text book of neurology or internal medicine, seems to us superfluous.

In regard to the **duration** and **frequency** of the attacks, everything that is essential has already been stated. It should be added that in the genuine form the attacks may in the beginning be nocturnal and, if the initial cry is absent, as is so frequently the case, the attacks may not be noticed for some time, or only inferred from the evacuation of urine by which they are accompanied (compare page 374).

Petit mal in its various forms is perhaps even more frequent in children than in adults. It may take the form of momentary loss of consciousness (absence), epileptic vertigo, or rudimentary, *i. e.*, atypical convulsions.

Dazed conditions and vertigo manifest themselves as in the adult by loss of consciousness (total analgesia!) with slight symptoms of motor irritation. In cases of absence the latter may be entirely absent, or consist merely in a slow, but very forcible rotation of the head, distortion of the face, or rolling of the eyes. In epileptic vertigo the patient usually falls to the ground and has a very short, tonic convulsion, often accompanied by involuntary evacuation of urine. The vertigo is often preceded by an aura.

In many children consciousness is merely clouded instead of being completely suspended during epileptic vertigo. They continue with what they are doing, and respond when spoken to, but they do not remember afterwards what happened during the attack.

Of the rudimentary and atypical attacks, the most important are: (1) localized twitchings, designated "secousses" (Herpin) and resembling those which occur at the beginning of the cortico-epileptic attack; (2) nodding epilepsy (epilepsia nutans); and (3) epilepsia procursiva.

The two first-named varieties, which are often unaccompanied by loss of consciousness, are always of short duration and are therefore rarely observed by the physician unless, as occasionally happens, the attacks are massed. It is difficult to identify them by the description obtained from parents. From the diagnostic standpoint it is important to remember that they may be an expression of a focal disease of the cortex (*Jacksonian epilepsy*) and that they also occur in genuine cases.

The attacks assume a variety of forms; sometimes they resemble the movements seen in tic; at others, a short lightning-like start with a jerking, forward movement of the trunk and head while the arms are extended, during which the child, if it happens to be sitting at the table, may strike its forehead against its plate.

Another form is the so-called *nodding epilepsy* (salaam spasm), in which the same movement, not unlike a bow, is repeated with lightning-like rapidity twenty, fifty, or even a hundred times in rapid succession. This variety of spasm must under all circumstances, be sharply distinguished from nodding spasm (see page 322), even when it is not accompanied by loss of consciousness and disturbances of the eye-movements are present also. On the other hand, there are cases which clinically resemble salaam spasm and which have nothing whatever to do with epilepsy; they must be regarded as cases of generalized tic, or as a hysterical symptom (hysterical salaam spasm).

A positive **diagnosis** of epilepsy demands either absolute proof that consciousness was clouded during the attack, or the presence of other positive epileptic phenomena in the same individual.

The term *epilepsia procursiva* has been used to describe certain peculiar epileptic attacks, consisting in forced running which begins suddenly and, after persisting for a variable time, terminates as suddenly, or ends in an attack of *grand mal*, thus revealing its true character of a motor aura. Consciousness is not always completely abolished during running, as appears from the fact that many of the patients avoid obstacles or turn about face when they come to the obstacle, etc.

Both nodding epilepsy and epilepsia procursiva appear to be rare. Personally I have never seen any cases.

Alterations in the psychic functions, in the form of either abolition or impairment of consciousness, or of a change in the individual's character, are practically constant accompaniments of the motor discharge. In addition, it has been known since the time of Esquirol that there may be paroxysmal psychic disturbances ("psychische equivalente," Hoffmann) which occur spontaneously and may alternate with the motor attacks. Typical examples of this *psychic epilepsy* are seen in the auræ (compare page 361), in ambulatory automatism, and certain psychotic conditions of short duration with maniacal excitement or depression. What has been already said in connection with epileptic auræ applies to all phenomena of this kind. They are much more rare in childhood than after puberty, and appear to occur only in genuine epilepsy. They are characterized by their sudden onset and disappearance, and by the utter failure to remember anything that occurred or that the patient has done during the time of the attack, which is in great contrast to the individual's behavior during the pyschotic change, which is often quite deliberate and orderly.

During the intervals of freedom many epileptic children exhibit cerebral—motor and psychic—disturbances which are termed collectively, intraparoxysmal symptoms. They are of considerable interest because they prove that epilepsy is a chronic, diffuse disease of the brain, but they have little diagnostic value because they develop comparatively late.

In our description of eclampsia infantum, we learned that mechanical and galvanic overexcitability of the peripheral nervous system is a necessary interparoxysmal symptom of the highest diagnostic importance, which is present from the beginning of the disease. The interparoxysmal symptoms of epilepsy are of a totally different nature. They are not latent symptoms of an abnormal condition of irritability which it is possible to discover by examining the patient, but rather afford clinical evidence that the epilepsy, or some other similar change in the brain has attained a considerable extent. In the main, therefore, they resemble the postparoxysmal symptoms of bodily and mental fatigue and exhaustion, and exhibit every grade of intensity. Lasting changes in the individual's character gradually develop. They may become ill-natured, silly, violent, quarrelsome, etc., and the intelligence may suffer to the point of pronounced epileptic idiocy.

These interparoxysmal symptoms develop gradually after the disease has lasted for a variable length of time, *i.e.*, when the periodical recurrence of the attacks has already established the diagnosis. They are of no assistance in interpreting the early, ambiguous attacks.

The cerebral symptoms of idiocy, infantile palsies, and the like, which precede or accompany epilepsy, or remain as permanent sequels of severe attacks, such, for example, as scars on the tongue, cutaneous hæmorrhage and the like, do not properly belong to this category.

In the **etiology** of epilepsy heredity plays an important part, particularly *direct homologous inheritance.* Sometimes the disease skips a generation so that, for example, epileptic children may have a healthy father and an epileptic grandfather. Inheritance from both parents, however, is only found in a small percentage of epileptic cases and, according to Marie's investigation, is much less frequent as an etiologic factor than some infectious disease. Without going into the numerous and contradictory statistics, we may say this much, that the frequency of inheritance from both parents is noticeable only in genuine epilepsy, beginning shortly before or at the time of puberty; while in symptomatic epilepsy it is of slight importance. In the latter variety of the disease, *transforming inheritance* and a general neuropathic taint, play a much more important, but at the same time unintelligible rôle, so that other etiologic factors appear to be more weighty. Among the latter are intra- or extra-uterine diseases of an infectious or toxic nature and, as exciting factors, physical and psychic injuries (fright).

Syphilis by producing a specific cerebral lesion may be the cause of symptomatic epilepsy. Other signs of focal disease besides the convulsions are practically always found in such cases. In others, which appear to be cases of genuine epilepsy, congenital syphilis probably has the same etiologic importance as any constitutional injury that affects the central nervous system, such as various intoxications, alcohol, lead and the like.

The relation existing between epilepsy and eclampsia infantum calls for special discussion.

Since it may be regarded as certain that the two diseases are fundamentally different, the transition from eclampsia to epilepsy is no more conceivable than a transition, for example, from hysteria to epilepsy. The question is whether eclamptic patients, or those who have suffered from eclampsia, possess a certain predisposition to epilepsy. The statistics which we find in the literature (see Finckh's analysis), and which without exception answer this question in the affirmative, are marred by two serious defects. In the first place, genuine and symptomatic epilepsy are not differentiated with sufficient accuracy; and in the second place, it is impossible, when a history of convulsions in infancy is obtained, to secure sufficient proof that the convulsions were eclamptic in character.* The only way in which positive conclusions can be reached is by keeping a series of eclamptic children under observation for a period of years instead of depending on retrospective examinations of epileptics, but this method has never been systematically employed. My own numerous observations, although they have not as yet been carried on for a sufficient length of time, tend to show that eclampsia does not indicate any predisposition to epilepsy. By this I do not mean to deny that a child who has had eclampsia may later become epileptic; but such cases are rare in comparison with the frequency of eclampsia in childhood.

When the case is merely reported by a layman and has not been under accurate medical observation, it is always possible that the first attacks during infancy may have been epileptic: or conversely, attacks occurring in the second or third period of childhood may be attacks of late eclampsia (compare page 343).

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* The unreliability of clinical data obtained from parents was forcibly borne in upon me while I was school inspector. I found that many parents in filling out a medical inspector's form answered the question as to whether the child had ever had convulsions in the negative, although I myself, as assistant in the children's clinic, had treated the same child for eclamptic convulsions. Such convulsions often make very little impression on the parents, and are only remembered when the child again develops convulsions at some later period

Quite independently of the data obtained by questioning the patient, which we have used as a starting point, we may speculate theoretically whether the convulsive attack itself, judged by its severity or frequent recurrence, may have been capable of producing the epileptic changes in the brain indirectly through the circulatory disturbances to which it may have given rise. Although this possibility cannot be altogether denied, there are so many well-founded objections to (for the literature see Freud) this view that its adherents are constantly diminishing in numbers.

Many authors see in *autointoxications* of the organism an important etiologic factor of epilepsy. As yet this doctrine of autointoxication is based on such a small number of authentic cases that we need not discuss it here and may refer the reader to the text books on epilepsy.

The same is true of the rare *reflex epilepsies*, the study of which must be begun all over again, taking into account what we now know of the spasmophile diathesis.

The **diagnosis** of epilepsy in its well-developed forms is easy when the chronic character of the disease has become manifest by the periodical recurrence of the motor or psychic attacks at intervals of months or years, and permanent interparoxysmal changes have perhaps already developed. It is much more difficult, and frequently quite impossible, on the other hand, to diagnose epilepsy from the first convulsive phenomena or psychic equivalents as observed by the patient's friends or family. The clinical expression of this emotional or inhibitory explosion is not in itself characteristic of epilepsy, and becomes so only by the periodical recurrence of the attacks.

In the present state of our knowledge, we are unable in every individual case, even after a careful examination of the patient, to decide whether a convulsive attack represents the first manifestation of an organic disease of the brain or merely a functional disturbance. There are functional convulsions the origin of which is absolutely unintelligible—at least none of the well-known causes, such as uræmia, for example, can be discovered—and such attacks we are inclined to regard as the first manifestations of epilepsy. In the subsequent course of these cases, however, we are often surprised to find that the convulsions remain limited to one or a very few attacks, and recur either not at all, or only after the lapse of many years.

In other cases, epilepsy can be excluded with more or less certainty after the first examination, and some other positive diagnosis can be made in its stead.

Aside from those cases in which the symptomatic nature of the convulsion at once becomes evident from the long duration (several hours) of the first attack, the attending high fever or other phenomena, there are in the main two diseases that have to be differentiated—"late eclampsia" and hysteria.

By the term "late eclampsia" I mean convulsions developing on the foundation of the spasmophile diathesis and differing from the ordinary eclamptic attacks of early childhood only by the greater age of the individual attacked. They usually, but not always, represent repetitions of the infantile convulsions. They simulate epilepsy chiefly in that they occur during the fifth to the eighth year of life (possibly somewhat later) and differ from eclampsia by the presence of pronounced tetanoid symptoms and their favorable prognosis.

Hysterical convulsions and dazed conditions are not rare in older children, that is, at the period of life when epilepsy frequently begins. If the physician has an opportunity of observing the attack himself, the absence of pupillary rigidity, of analgesia and other characteristic symptoms will usually enable him to arrive at a correct diagnosis; but a hysterical child is not always willing to perform for the physician's benefit, and even in the hospital the physician is not always so fortunate as to witness the attack.

In such cases the differential diagnosis, aside from the general considerations discussed in the chapter on Hysteria, must be made by the aid of the following special points:—

If the attack is violent and comes on suddenly, if the patient is injured during the attack, and urine and feces are evacuated involuntarily; if it is followed by prolonged sleep and the patient has no recollection of its occurrence, epilepsy is the more probable diagnosis. Uniformity, short duration, and recurrence at approximately regular intervals of a few weeks, also point to the diagnosis of epilepsy.

On the other hand, if the conditions are exactly opposite with respect to the above-mentioned points, it is a much less powerful argument in favor of hysteria. It is only when the patient distinctly remembers the attack when it is brought on by emotion, and suggestive treatment is followed by permanent recovery that the diagnosis of hysteria can be made with any degree of certainty. We would especially warn against overestimating the significance of the temporary disappearance of the attacks under suggestive treatment; for a temporary cure of this kind may be simulated by what is the result of pure accident in cases which are undoubtedly epileptic.

The **prognosis** of epilepsy is always exceedingly grave. Aside from the fact that the patient may die in the attack as the result of some accident or from exhaustion, it is rarely possible to prevent recurrence of the attacks and the gradual development of the post-epileptic physical and mental decay. The diagnosis must be questioned in all cases with rapid recovery.

The treatment of epilepsy is a thankless task, even if it is instituted very soon after the first attacks. As the treatment is the same in childhood as in adults, we shall merely touch upon the most important points.

Treatment.—The epileptic attack itself usually calls for no special treatment, except to guard the child from injury during the convulsions, from biting its tongue, and from aspiration of the tongue, by drawing the member forward. If there is high fever, wet packs may be tried. If the attack is abnormally protracted and in the status epilepticus chloral in doses of 0.5 to 1.0 to 2.0 Gm. (8 to 15 to 30 grains) per rectum, or inhalations of chloroform act as a sufficient sedative.

To prevent recurrence of the attack the child should be placed under the best hygienic surroundings that can be achieved with regard to diet, sleep, and light mental occupation under proper supervision; and in addition, medicinal treatment must be begun at once. If there is a possibility of the epilepsy being of the symptomatic type, and even in the absence of any suspicion of syphilis, a vigorous course of treatment with mercury and potassium iodide should be tried because of their known favorable effects on inflammatory brain processes. If, however, no results are obtained, the treatment must not be kept up longer than from four to six weeks. The administration of bromides must then be begun. The potassium salt, or Erlenmeyer's bromide mixture (potassium bromide and sodium bromide, of each 1.0 Gm. (15 grains), ammonium bromide 0.5 Gm. (8 grains), or one of the newer organic bromide combinations may be selected. Whether gradually ascending or descending doses should be prescribed, or a constant dose given for a considerable period appears to be a matter of taste.

The important point is not to be satisfied with small doses of bromides if no result is obtained. The attacks must be suppressed, if necessary by the exhibition of as much as 3 to 5 to 6 Gm. (45 to 60 to 90 gr.) per day, to a child of five years, and 8 to 10 to 12 Gm. (2 to 2½ to 3 drams) to a child of ten years. Under medical supervision there is no danger in doses of this size, and the disturbances collectively known as "bromism" can, if necessary, be made to disappear rapidly by withdrawing the drug for a time. If a cure, or only a moderate improvement in the severity of the epileptic manifestations is effected by the bromide treatment, it should be continued indefinitely, possibly with occasional short interruptions. This is the unanimous opinion of all competent physicians.

In order to reinforce the effect of the bromides and enable the patient to get along with as small a quantity as possible, when it is to be taken for a long time, we may adopt the suggestion of Richet and Toulouse to keep the patient on a diet containing as little salt as possible.

If very little or no effect at all is produced by the bromides, belladonna or a combination of bromides and opium, after Flechsig, may be tried, although but little is to be hoped from this method.

The question of surgical interference rarely comes up in the case of children. The indications are the same as in adults.

Many of these unfortunates end their lives in institutions for the care of epileptics.

NEURASTHENIA

Since we have become familiar with the neurasthenia of adults not only in its severe and well-developed forms but also in its initial and milder, rudimentary manifestations, there can be no doubt that children are subject to neurasthenia.

The exact nature of the functional anomaly which we now call neurasthenia is best described by the words "irritable weakness," *i. e.*, a pathologic increase of irritability and diminished resistance to fatigue. The two factors go hand in hand, but not in the same degree, the irritability being sometimes more marked, while at other times weakness is the most prominent factor.

Although neurasthenia represents a disease of the entire nervous system, we do not find in an individual case that all the various nervous functions are equally affected. Quite often the disturbances appear to be isolated or localized, so to speak, and it is only after a thorough examination of the patient, or more frequently after we have observed his subsequent development, that we realize that the individual symptom is the expression of a pathologic change which is responsible for a variety of other symptoms. This explains the variegated character of the disease.

Before describing the symptomatology, a few words must be devoted to a general question, namely, the distinction between neurasthenia and hysteria. While the two diseases are frequently combined in the same individual, nevertheless, since we have adopted Möbius' law* in regard to the psychogenic origin of hysterical symptoms we refuse to recognize as genuine transitional forms either "neuro-hysteria" or "neuro-epilepsy." In our opinion these conditions merely represent combinations—intricate and difficult to unravel, it is true of two related, but nevertheless intrinsically distinct neuroses, neurasthenia and hysteria. At the same time we can readily see that the same clinical symptoms may be produced by either of the two neuroses, as will be illustrated by examples both in this chapter and in that on Hysteria.

Neurasthenia is by no means always a permanent, invariable, constitutional anomaly; it may be temporary, developing under the influence of bodily or psychic fatigue, and disappear as soon as the cause is removed; but there is no essential difference in the manifestations of the two forms.

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* See chapter on Hysteria.

Symptomatology.—Unlike hysteria, the occurrence of neurasthenic symptoms does not depend on a certain degree of psychic development, and we accordingly see the abnormal irritability manifesting itself in the simple reflex processes of very young infants. Perhaps the earliest symptom is an abnormal tendency to fright; the sudden starting of the sleeping or quietly resting infant under the influence of a sudden noise, or bright illumination. The sudden start is sometimes accompanied by a cry, which is purely reflex; it has as yet no psychic counterpart, there is no fear of any impending evil. The violent crying and struggling of many young infants in the bathtub is a similar phenomenon. The mothers usually think the child is afraid of falling. Whether the child is distressed by vertigo or some similar sensation, we do not know. Continuous crying, day and night, and abnormally light sleep may also be early symptoms of neurasthenia; but one should be cautious in making such a diagnosis, because somatic diseases (disturbances of nutrition, interference with nasal respiration from coryza) may be the true cause. Sometimes mild general twitching of the whole body can be elicited by tapping any part of the person. It is very difficult to estimate whether the reflexes are exaggerated in infants.

The symptoms of neurasthenia are both somatic and psychic. Among the former we distinguish between *subjective* and *paroxysmal*, such as headache, vomiting, syncope; and the *objective* and *permanent* symptoms, which are comparable to the stigmata of hysteria and of great diagnostic value because they can always be discovered by examining the patient. It should be emphasized, however, that they are frequently absent or present only in part in cases of pronounced neurasthenia. These objective and permanent symptoms are: Increase of the tendon and periosteal reflexes, the cutaneous and mucous membrane reflexes being normal or greatly diminished (absence of corneal reflex and pharyngeal reflexes); tremor of the eyelids, or incomplete closure of the palpebral fissure when the child attempts to close its eyes (Rosenbach's phenomenon); increase in blood pressure* to 110 or over 150 mg. of mercury instead of 80 to 90 mg. (Heim, Strauss); changes in the pulse rate; alternating inequality of the pupils (Mikloszewski, Pick, Schoumann and others); evanescent variable differences in the innervation of the two sides of the body, observed both in the facial muscles and in the muscles of the extremities; sudden changes of color and the appearance of fleeting erythema in the temples, neck, upper portions of the chest (erythema pudendi) when the clothing is removed for purposes of examination; slight tremor and shaking of the hands; hurried, noisy manner of speech or stammering. Anæsthesia such as occurs in hysteria does not belong to the picture of neurasthenia, and the above-mentioned absence of pharyngeal and corneal reflexes is by no means accompanied by anæsthesia or hyperæsthesia of these parts (Kiewe). Similarly Bartenstein has shown that Head's zones in children cannot be regarded as stigmata of neurasthenia in the absence of disease of the internal organs.

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* Measured with Gärtner's tonometer.

The above-mentioned symptoms have very different diagnostic values; the presence of one alone proves absolutely nothing. It must be remembered also that many of them may have an organic cause—as for example, increase in the reflexes; abnormal increase of the blood pressure (inflammation of the kidneys); inequality of the pupils (enlargement of the bronchial and mediastinal glands stimulating the dilator fibres in the sympathetic nerve); tremor and the like. Others, such as stammering, may represent an independent disease; others again, such as Rosenbach's symptom, tremor of the hands, among others, are extremely elusive, they are indications of uneasiness and excitement rather than signs of the neuropathic constitution and for that reason observed only at the first examination, after which the child ceases to be afraid of the doctor. It should also be stated that the facial phenomenon (Frankl-Hochwart and others), which is often included among the symptoms of neurasthenia is, in our opinion, to be regarded not as a general nervous symptom but as a characteristic sign of latent spasmophilia (Thiemich) (compare page 317).

The **paroxysmal somatic manifestations** of infantile neurasthenia are many and various. Often they show themselves only under special circumstances, such as the presence of some acute general disease, because the normal power of inhibition is absent and certain symptoms on that account assume alarming dimensions and exhibit an abnormal degree of severity. These are seen in diseases of the respiratory organs which are accompanied by cough. Where a child with sound nerves would merely have an ordinary cough, a nervous child will develop convulsive cough of alarming severity, with cyanosis, choking and vomiting. There is no doubt that cases of this kind are often mistaken for whooping-cough, even by physicians. Vomiting is a neuropathic symptom, produced in many children by any kind of excitement—for instance, when they are awakened in the morning to go to school, or during the walk to school, or as soon as they have entered the classroom. Constipation also occurs in children as in adult "hypochondriacs," although diarrhœa, or even incontinence of feces, is more frequent as the result of worry or excitement. These, however, are graver forms of the disease. Loss of appetite is a frequent symptom.

Diurnal and nocturnal enuresis and pollakiuria, which are usually due to a general neuropathic constitution, have been sufficiently discussed in the chapter on Hysteria (see page 373). In this connection mention should also be made of the inability to urinate in the presence of other persons—even near relatives, which is observed in many children.

Palpitation and arrhythymia are also symptoms of neurasthenia. The neuropathic symptoms referable to the nervous system, such as vertigo on looking down from a great height or riding backwards in a carriage, or under other similar circumstances, sudden syncope from fright, fatigue and like causes, occupy an intermediate position between the somatic and the purely psychic phenomena. Mention must also be made of *nervous headache,* which is one of the most frequent somatic symptoms in neuropathic children. As a rule it does not appear until the child is old enough to go to school, and of all the neuropathic symptoms is probably the one that is the most frequently attributable to the stress of school work. Forced attention, not the result of spontaneous interest in the subject under discussion, a restless anxiety as to how the lessons will turn out, or positive fear when the lesson has not been properly prepared or understood; in short, the whole array of school worries, which adults often smile at so inconsiderately, may produce headache. Other things that contribute to the headache are the enforced immobility for several hours and the vitiated and overheated air which accumulates in an ill-ventilated class-room by the end of the hour, particularly as these children are usually pale and flabby and habitually suffer from cold hands and feet, or, in other words, from improper distribution of the blood. Hence we are often told, and we have no reason to disbelieve the statement, that the headache readily disappears during the holidays and returns a week or two after school begins.

The headache may be present in the morning, but usually does not develop until after several hours of lessons, sometimes earlier sometimes later, disappearing gradually during the noonday meal or persisting until the child goes to bed. It does not, as a rule, appear to be very severe; the child rarely wants to lie down unless accustomed to do so by the anxious mother. It is usually described as a dull headache, chiefly frontal; the variety described as affecting the entire skull like an ill-fitting helmet (casque neurasthénique, Charcot) seems to be rare in children. The scalp is often sensitive, sometimes so much so that combing the hair causes intense pain and is given up on that account. Although this "school headache" which we have just described is usually neurasthenic, there are also many cases in which it proves to be an hysterical imitation. Thus as a school inspector I would find that in some classes of girls a third or more of the children, usually between the ages of 10 to 14, when questioned, would answer that they had headache; whereas, in another class of 40 to 50 girls of the same age, only a few complained of headache. The oftener the question is asked during the year in the same class, the greater will be found the number of children complaining of headache. When large numbers of these children are treated by suggestion, giving them daily a few drops of bitter tincture, or compound tincture of cinchona, or a short douche to the forehead, a rapid and permanent cure is effected in a great many cases (Herrmann). Such cases are hysterical, whereas the same treatment is without effect in neurasthenic children.

Another typical neurasthenic complaint is *nervous asthenopia*, which has been exhaustively studied by Wilbrand and Sänger and which most commonly develops between the ages of 10 and 14.

"The patients complain of flickering before the eyes; they say that the letters and lines on a page disappear as they look at them, that everything is confused, and that reading makes the eyes water and gives them violent pain in the brow and eyes. When a child complaining of these symptoms is asked to read aloud it is noticed that it soon begins to halt; the individual words are not properly recognized, and sometimes whole syllables are improperly pronounced. The child gradually holds the book nearer and nearer; for a time it is able to read a few words correctly, and then the old trouble begins again. Finally, when the little patient is holding the book so close that it almost touches its nose, the reading stops altogether and a series of peculiar manœuvres, rotation of the head, turning of the book toward the source of light, and so on, are begun without producing any improvement in the vision. These visible efforts to read are accompanied by wrinkling of the forehead, contraction of the eyebrows and of the orbicularis palpebrarum, twitching of the lips, and in short, by a series of active associated movements in the entire muscular region supplied by the facial nerve.

"According to Wilbrand and Sänger, this affection is quite frequent; but although we paid especial attention to the matter we hardly ever found a case of it in our own material. Nevertheless, it is of some importance as a typical neurasthenic affection.

"With regard to the psychic behavior of the child, the irritable weakness first manifests itself in a striking uncertainty of disposition, while the intelligence, on the other hand, seems to be but little affected. Owing to the diminished resistance to fatigue, the child's attention soon flags and it loses interest in studies which it had previously pursued with pleasure and a desire to excel; so that children who are suffering from neurasthenia, although of normal intelligence, go on from bad to worse in their school work and exhibit a constantly increasing dulness (neurasthenia cerebralis, Emminghaus). This dulness is often the expression of a melancholic or hypochondriacal depression, which also is the direct result of the subjective sense of weakness.

"In young neurasthenic children fear is the most prominent psychic symptom. The children are afraid of everything strange that they are brought in contact with; they are afraid of any friendly little dog or cat, of a new toy, of a strange face or unfamiliar room. At the same time this fear, which manifests itself in wild outcries and struggles and a terrified expression of countenance, has no normal motive and is not due to any past experience of harm: for even a child with sound nerves will cry out on seeing a dog, if it has ever been bitten, and bawls and struggles against the doctor, if the latter has ever hurt him. Older children usually control themselves to the extent of suppressing their tears and manage to keep from crying out, but it is easy to see that fear is gripping them by the throat and that they are hardly able to speak. At the same time they remain at a respectful distance from the doctor and tremble whenever he makes the slightest movement.

"Sometimes the children hide their fears under an assumption of courage or obstinacy. Thus, I know a four-year-old neurasthenic who, whenever he is spoken to by a stranger, shakes his fist at him, although with a frightened expression of countenance, and threatens to "soak him one," but at once bursts into tears and tries to run away if one attempts to molest him.

"Fear as well as joy, especially in older children, finds expression in severe paroxysms of sustained convulsive laughing or crying. Children who as soon as they enter the consulting room, or at the latest when they are undressed, make a wild scene, and throw themselves on the floor in a fit of rage, are always neurasthenic children that have also been badly brought up. In many of these neurasthenic children the emotion of fear or anger, sometimes also of sudden fright after some inadvertent injury, produces brief attacks of a dazed condition ('*Wegbleiben*') which the laity quite aptly describe as 'convulsive rage.'

"These attacks have recently been described in detail by Neumann. When the child attempts to cry it is unable to progress beyond the stage of inspiration; the expiratory muscles remain in spasmodic contraction and breathing is arrested. In desperation the child strikes out with its fists once or twice, the face becomes pale and the entire body rigid, the eyes are turned up, and consciousness is lost. As a rule, involuntary evacuation of the bowels does not take place. Usually the attack lasts only a few seconds, although to the anxious relatives it often seems as many minutes. As the spasm relaxes the child, who in the meantime has usually fallen backward, rarely forward, at last manages to catch its breath and gives a loud cry, which may or may not be preceded by a short inspiration, as if the child had at last succeeded in making the intended expiratory movement which it was prevented from effecting by the spasm of the inspiratory muscles."

As appears from this description, attacks of this kind may show a very great similarity to the expiratory apnœa which is related to laryngospasm (see page 308).

The **diagnosis** is made by the regular absence of inspiratory stridor and of galvanic overexcitability of the nervous system, and these attacks of convulsive rage cannot therefore be included among the phenomena of spasmophilia. There is also a difference in the age of the children. Whereas spasmophile convulsions occur chiefly between the ages of six and eighteen months, the above-described attacks of convulsive rage usually do not manifest themselves until the second or third year of life and almost always disappear at the latest when the child reaches the age of five. An attack can often be prevented by educational measures, by making it clear to the child that any attempt to hold its breath will be immediately followed by severe punishment; in this way one may often succeed in preventing the spasm by a blow or by merely threatening to strike. Once the attack has developed it can be shortened by dashing cold water on the face, by a light blow or some other cutaneous irritant; but even without interference of any kind no fear need be entertained for the child's life or any other injury, aside from the possibility of its hurting itself when it falls to the ground, and in this respect the attack differs essentially from spasmophile convulsions.

Disturbances of sleep occur in association with a great variety of diseases in childhood, as well as among adults, and indicate pain, fever or some other disturbance. In the absence of any physical basis we are justified in regarding the disturbance as a nervous phenomenon; in fact, objective investigation and observation of children whose sleep is habitually disturbed have shown that these children are nervous individuals and that the troubled sleep is merely a symptom of the constitutional anomaly.

The *clinical manifestations* are various. Some children, both infants and older children, instead of going to sleep at once after they have been sufficiently fed and put to bed, as normal children do, lie awake for hours, but when sleep finally comes on it is of normal depth and duration. Some children will not go to sleep unless they are allowed to suck a finger or a corner of the bedclothes; others must have the face completely covered with the bedclothes; and some cannot sleep unless they are sitting up or lying on their stomachs. These are mere habits for which the children's surroundings are frequently responsible, but the rapidity and obstinacy with which these habits become established nevertheless indicate the presence of some pathologic factor in the child's makeup. There are other cases in which the children go to sleep promptly, but do not sleep soundly and are easily aroused by a show of light, or even without any recognizable cause. Some children exhibit a variety of motor phenomena, such as grinding the teeth, rolling about, flexion and extension of the arms and legs, and the like, which indicate that they are not sleeping soundly. These movements continue without wakening the child and may be rhythmical and quite violent, presenting the character of stereotypias (jactatio capitis nocturna) [Swoboda, Zappert].

Many children as they drop off to sleep go through complicated actions which suggest dreams. They are apparently playing with some toy, or occupied with their school duties, point upward with the finger and indulge in other similar antics.

A good many children during the school age are given to *somnambulism;* they get out of bed, go from the bedroom, for example, to the nursery, play with their books, and finally return to bed unless they are seen by some one and wakened. The child's behavior in the somnambulistic state is perfectly quiet, and it is quite unconscious, in contradistinction to its behavior during attacks of night terror, pavor nocturnus, which we shall now proceed to describe.

Pavor nocturnus, or night terror, is a special form of nervous disturbance which in the literature is usually described as an independent affection, although many authors have recognized, and insisted upon its dependence on a nervous constitution.

Night terror occurs most frequently between the ages of three and six years, but may persist in older children up to the beginning of puberty. Both sexes are equally affected. The attacks usually occur one or several hours after the child has gone to sleep. It wakes up with a cry, sits up in bed, and manifests other signs of extreme terror, which often seem to be due to some terrible dream, judging from the words and fragments of sentences which the child utters during the attack. In a terrified manner it begs the nurse to keep off the dog or man that is frightening it, to hold on to it, not to let it drop, not to punish it, etc. Even after the light is turned up and the mother has taken the child up to comfort it, the terror still continues, and it takes from a quarter to half an hour before the child recognizes the situation and allows itself to be pacified, and goes to sleep again. Sometimes the attack recurs in a milder form once or rarely twice in the same night. As a rule, the child remembers nothing of the attack on awakening in the morning.

The intensity as well as the frequency of the attacks is extremely variable; sometimes attacks of day terror—*pavor diurnus*—occur in the daytime.

A variety of physical causes have been suggested for the occurrence of the attacks, such as worms, digestive disturbances, overloading of the stomach, constipation and the like. Rey called attention to the presence of adenoid vegetations in many cases. He believes that the interference with respiration and the consequent accumulation of carbon dioxide causes intoxication of the brain and a nervous discharge in the form of an attack. These possible causes must of course receive attention in the treatment of this class of patients; it is to be remembered, however, that they are absent in a great many cases of night terror and, conversely, that they are present in many children without producing any attacks. The increased susceptibility to carbon dioxide accumulation, which is mentioned by Rey, is after all but a sign of an abnormally irritable nervous system.

Neither somnambulism nor night terror has anything to do with epilepsy.

In connection with this nocturnal disturbance it should be cursorily mentioned that some children are very difficult to wake, even after a long night's rest. In the morning they are drunk with sleep, irritable when they are called, and continue to be half awake while they are dressing and eating their breakfast. It takes these children a long time to wake up completely. In psychopathic institutions, particularly, it is often noticed that the more nervous children do not accomplish as much during the first hour as they do later in the day.

Heredity plays an essential part in the **etiology** of neurasthenia. It is, however, usually associated with injurious environment and improper training. An irregular mode of life, restlessness and dissensions at home, being allowed to take part in the amusements of adults, usually at the expense of sleep, and the reading of books that excite the imagination, etc., greatly undermine the child's nervous constitution. The mental strain of school work, particularly in children who are naturally nervous and not very highly gifted intellectually, and who are made to begin school too early and forced in their studies, is also a potent factor of evil.

A severe somatic disease, as well as frequently recurring milder illnesses (as, for example, in children with a tendency to exudative diseases) may give rise to neurasthenic conditions; but it is a mistake in our opinion, to say that every child of school-going age that looks a little pale is anæmic, and to accept that as sufficient explanation for existing nervous symptoms.

Finally, the bad effects on the child's nervous system of indulging regularly in alcoholic beverages, must be alluded to, although this very modern theme requires no further elaboration.

The **diagnosis** of neurasthenia is not difficult in most cases, but the child must always be subjected to a searching examination for the existence of any organic disease that may be either the cause, or merely a concomitant of the neuropathic symptoms, which are the most prominent features of the clinical picture. Many cerebral affections, such as tumors, epilepsy and chorea, for a long time give rise only to general neuropathic symptoms, until finally the grave characteristic signs of the organic disease make their appearance.

The **prognosis** of infantile neurasthenia depends partly on the severity of the inherited disability and partly on the environment, whether favorable or unfavorable to the child's development. Under favorable conditions complete recovery is possible.

In many cases the most important part of the **treatment** is the removal of the most prominent symptoms, which may be accomplished by the usual methods employed with a certain measure of discretion. The conscientious physician, however, will never lose sight of the causative disease and will therefore scrupulously avoid polypragmasia and a mere pretense at treatment for the sake of doing something. The egotism and the tendency to hypochondriacal self-analysis, which are characteristic of neurasthenic children—astonishing examples of this tendency are often seen in children of a tender age—are in danger of being enhanced by constant medical treatment, both directly and indirectly, and by the constant attention such children receive from their parents. Indeed, hysteria may be directly produced by such a policy. Besides, drugs at this age usually have but little effect and may be dispensed with altogether. This is especially true of the great multitude of nutrients, tonics, hæmatonics and other remedies which are shamelessly advertised and placed on the market by our modern industrial institutions.

In almost every instance the symptoms can be made to disappear by correcting faulty environment, by restricting well-meant but ill-advised educational methods, and by regulating the child's mode of life, providing it is in the physician's power to do so.

As an example, we may mention the symptom of *anorexia*. Every physician is familiar with the type of nervous child that never asks for anything to eat and for whom every meal, especially the principal meal of the day, is a source of terror lest the anxious, neuropathic father may force it to eat by threatening a flogging. Every physician knows also that he can accomplish more by forbidding every form of coercion than with any medicine or diet-list that he might order, and the children of this type, when they are away for their summer holidays or on a visit to relatives in the country, learn to eat without any trouble. It is equally irrational to put a neuropathic child on a forced diet that is quite beyond its appetite, unless it is at the same time placed in a sanatorium where it will be separated from fussy parents who unconsciously create an amosphere of excitement and unrest—a plan which must always be borne in mind as a last resort.

HEREDITARY NEUROPATHIA

(HEREDITARY DEGENERATE PSYCHOPATHIC CONSTITUTION)

Under this head we group a number of morbid phenomena which, although they are frequently included in the broad, elastic definition of nervousness, cannot be called neurasthenia in the sense used above, but represent severer neuropathic anomalies and with gradual transitions to the true psychoses.

Under this head we classify migraine, tic, the various forms of stereotypia and the phobias, uncontrollable ideas and actions, pathologic dreaming, the travelling mania (dromomania) excessive masturbation and, finally, suicide in childhood.

It is evident that these morbid conditions only roughly correspond with what is frequently described in the literature as psychasthenia, psychopathic deficiency, or—to use the most comprehensive phrase—abnormal phenomena in the psychic life of children.

The significance of these symptoms is exceedingly variable, depending on whether they are associated with diminution of the intelligence or of the ethical sense, or, as in other cases, with unusual gifts greatly exceeding the average. An attempt has been made to take account of these peculiar differences by sub-dividing degenerates into "inferior" and "superior" degenerates.

It must be pointed out that many of these conditions present a very close, although as yet but little understood relation to genuine epilepsy. Indeed they might properly be discussed under the same heading, and in devoting a separate chapter to epilepsy we have been swayed chiefly by practical considerations.

MIGRAINE (HEMICRANIA)

Migraine presents a form of hereditary degeneracy, the subjects of which are classified among the superior degenerates; in other words, the disease does not represent a more or less uniform, disabling degeneration affecting the entire personality, but rather a practically isolated affection. It is well known that many persons of unusual mental gifts have suffered from migraine.

Migraine manifests itself in periodic attacks which, while they may vary in certain respects, all have as a characteristic feature the occurrence of more or less exclusively unilateral paræsthesias presumably due to cerebral processes. A complete attack is composed of an aura, followed by headache and vomiting. Instances in which the attacks are incomplete or partial are, however, more common. This very incompleteness of the attack often makes it difficult to recognize the disease as migraine, and if, with Henoch and H. Neumann, we admit that the headache need not necessarily be unilateral and that in children it is usually localized in the forehead or even in the occiput, the dividing line between hemicrania and other forms of headache becomes more uncertain and arbitrary. The only characteristic feature we then have left is the periodic return of violent headache, more or less constantly attended by nausea and vomiting and followed by sleep lasting several hours, after which the patient feels perfectly well.

The uncertainty of the *diagnosis* is no doubt responsible for the marked discrepancies in the literature in regard to the frequency of migraine in childhood. Oppenheim, for example, states in his Lehrbuch der Nervenkrankheiten that migraine usually begins at the period of puberty; while H. Neumann reports that in 9 out of 13 cases which he observed it occurred between the second and fifth year, in 21 between the sixth and tenth, and in 13 between the eleventh and fifteenth year of life.

According to our own experience, and the material in the Breslau Children's Hospital (Herrmann), hemicrania is exceedingly rare in childhood; but this may be due to local conditions. As migraine, probably more so than any other nervous disease, is produced by direct homologous inheritance, especially from the mother's side, we made a point, in taking the histories, to question the mothers in regard to their own previous diseases, and in innumerable instances were told that they had had "head cramp" and all kinds of atypical attacks of headache; but rarely obtained a clear description of unilateral pain.

In view of this state of affairs, we shall have to content ourselves with merely stating the conflicting opinions, leaving the distinction between migraine and nervous headache to individual judgment. In typical cases, such as undoubtedly occur in older children, as shown, for example, by the trustworthy reports of Sachs, the disease is in every respect so like the migraine of adults that we are justified in omitting a more detailed discussion.

TIC CONVULSIF

This term is applied to a peculiar disease which has been especially studied by two of Charcot's students, Gilles de la Tourette and G. Guinon, and is therefore often called after these two authors. The disturbance manifests itself in a variety of *stereotyped* muscular movements, recurring at regular intervals and always identical in character, in the same individual. The course is chronic.

A certain external resemblance which the disease exhibits to chorea minor induced Weir Mitchell to speak of a "habit chorea," but this term should be avoided because the disease has nothing whatever to do with chorea. The term "coördinating memory convulsions," selected by Friedreich, more correctly describes the nature of the malady, as will presently appear.

Facial tic is the simplest form of the disease. It consists of short, clonic contractions or twitchings in one, or rarely both sides of the face. As a rule it does not affect the entire distribution of the facial nerve, but rather resembles certain isolated movements of facial expression, such as blinking, wrinkling of the forehead, raising the eyebrows, showing the teeth, drawing up the corners of the mouth as in laughing, etc., repeated at short intervals.

The twitchings in the facial muscles may be isolated or may be combined with other tic movements.

In rarer cases they are the expression of a circumscribed cortical lesion and are then to be regarded as abortive forms of cortical epilepsy, or of a reflex irritation of the facial through the trigeminal—*tic douloureux*.

As a rule, however, the underlying condition is a psychomotor functional disturbance similar to that which exists in generalized tic.

The tic movements, as has been stated, are of all kinds, shaking, rotating, or nodding of the head, rotation or jerking of one or both shoulders, jerking movements of the arms or legs, grasping, stamping, hopping, jumping, climbing, and bicycle riding movements, of every conceivable variety. Some patients habitually pick their noses or their chins, and in almost every case tic movements are observed in the face.

The movements resemble those of chorea, but are as a rule more rapid and more forcible and, when first seen, give the observer more the impression of a conscious or volitional, purposive movement. For this reason the term coördinated tic is used. The movements, however, are characterized by absence of purpose, by their intensity and stereotypic repetition at variable intervals, or sometimes in series.

Frequently the muscles of articulation, phonation and respiration are also involved, giving rise to the production of inarticulate choking, clacking, or other animal-like sounds, spitting (like a cat), barking, and similar noises.

Sometimes there is an uncontrollable impulse to utter obscene words (koprolalia), although this appears to be rare in children; or meaningless concatenation of syllables, or a tendency to repeat over and over again (stereotypically) words accidentally overheard (echolalia).

When the patient is completely absorbed in some occupation, or if his attention can be distracted, the tic movements diminish in intensity or cease altogether, as during sleep. Excitement and the consciousness of being observed increase the movements. By a strong effort of the will tic can be controlled for a time, but at the expense of a painful sense of coercion.

Psychic factors are the most prominent in the **etiology**, and the disease is undoubtedly closely related to hysteria.

Many cases must be regarded as hysterical, particularly those which exhibit dancing, jumping, stamping and other similar movements, which have received the special name of saltatory reflex convulsions (Bamberger). Its close connection with astasia and abasia, which have been described among the hysterical symptoms is obvious. The knowledge of this pathogenic relationship is of great value from a therapeutic standpoint, because the above conditions are susceptible of rapid cure.

In other cases, if the opportunity is afforded to observe the development of the disease, "it is found that in a great majority the movement is a coördinated forced movement, which is at first performed voluntarily, usually to get rid of some unpleasant localized sensation, and that long after the object for which it was performed has ceased to exist, the movement continues and is repeated again and again in an involuntary and altogether automatic manner. The presence of a foreign body in the conjunctival sac or a phlyctenula first causes the child to blink, and this blinking movement, long after the foreign body has been removed or the phlyctenula has healed, persists as tic" (Pick). In many other cases the first beginning of the convulsive tic can be traced to an originally normal, purposive movement.

The *pathologic* feature of the process consists in the persistence of the movement as an automatic and forced phenomenon, which is much more pronounced than the subcortical character of movements which have been learned by practice—a process which is quite normal, particularly in children.

The development of the so-called *stereotypias* depends on a similar psychic mechanism leading to automatism. These movements, however, lack the convulsive character and are quite frequently observed in children who exhibit no signs of degeneracy. Among these we may mention biting of the nails, scratching the head, picking at the fingers and the lips, and sucking movements. Special forms of stereotypia occurring during sleep have been mentioned on page 351 (Swoboda, Zappert, and others). Similar rocking or turning movements, called pagoda movements, are often observed in imbecile children and may be kept up for hours in monotonous repetition; like the sucking movements in normal children, these movements are often accompanied by obvious signs of well-being. Perhaps this feeling of pleasure which appears to be produced by stereotype movements of this kind, and which may go on to a veritable orgasm, is responsible for the fact that these movements have been identified with the onanism of infants and young children.

The **prognosis** of tic, so far as recovery is concerned, was pronounced by Charcot and his followers to be practically unfavorable; but their pessimistic view is probably to be explained by the profoundly degenerate character of their clinical material. Pitres and others, on the other hand, reported a number of favorable results, and to-day we may say that some cases—which thereby reveal their hysterical character—are susceptible to suggestive treatment, and that many of the cases which are due to inherited degeneracy can be greatly improved or even cured by suitable educational treatment.

Treatment.—Starting with the most favorable assumption, namely, that the condition is due to hysteria, antihysteric measures (isolation, intentional neglect, etc.), should be instituted at once. If these measures fail, and the necessity for educational treatment becomes apparent, the child should be placed in a suitable institution, as that offers the only prospect of ultimate success. It is only by surrounding the child with influences calculated to improve its general psychic condition that we can hope for any results from special treatment of tic, either by respiratory gymnastics (after Pitres) or by the use of mirrors (after Brissaud).

PHOBIAS

In their pronounced forms the phobias are rarer in childhood than among adults; nevertheless a number of cases of agoraphobia, dread of clothing, dread of travelling, dread at the sight of sharp objects, etc., have been described. The diagnosis is usually easy, and the prognosis as unfavorable as that of all morbid phenomena that are due to inherited degeneracy. The treatment will be discussed later in connection with the latter. These phobias are frequently associated with

UNCONTROLLABLE IDEAS (OR CONCEPTIONS)

These apparently are the original causes of the phobias. Thus Oppenheim relates of a girl 10 years old, "who in early childhood was seized with violent attacks of fright when one of the family, especially her father or mother, left the house. The child would stand in the doorway or at the window, trembling with fear and excitement, and could not be induced to move until the parents returned. In the course of treatment the condition became worse, so that the mother could not even leave the room, and finally the child developed a permanent condition of fright which so completely dominated all her thoughts and actions that it resembled insanity in every respect. It required very searching investigation to determine that an uncontrollable idea, namely, that an accident might happen to one of her family, was at the bottom of her trouble and was the only cause for the child's peculiar behavior. There was an entire lack of characteristic features of illusions because, as soon as the paroxysm of fear had passed, the child was fully aware that nothing would happen to its parents, and, in fact, realized that her fear was morbid." After Oppenheim had recognized the nature of the disease he succeeded by suitable treatment in bringing about a considerable improvement in the child's condition.

This case illustrates the peculiarity of uncontrollable ideas, which was emphasized by Westphal, namely, that the idea is felt by the patient to be something unusual and morbid, although at the same time it cannot be suppressed either by an effort of the will or by corrective counter-ideas. In other words, the condition differs from illusion in that the patient is aware of his malady. This consciousness, however, as Pick has pointed out, may be absent in young children whose intelligence is not yet sufficiently developed, but such absence should not be taken as a sign of illusion or insanity.

In children, as in adults, uncontrollable ideas assume various forms, and some have a greater pathologic significance than others. As most children do not willingly reveal their inner lives, it often happens that the physician first hears of the existence of the forced ideas in the patient's childhood from the latter's own lips after he has grown up. In other cases *uncontrollable actions*, which are the result of uncontrollable ideas, lead to the early recognition of the abnormality.

The latter variety of forced or uncontrollable ideas is always to be regarded as pathologic, whereas many *milder forms*, such as an uncontrollable impulse to count the houses or lamp-posts, to step on the cracks of the pavement, and the like, appear to be quite common among children. Sometimes a child becomes aware of the meaninglessness of its thoughts and makes an attempt to break up the habit; sometimes the habit is begun as a mere pastime and is soon given up.

Of the many *graver forms* that occur in adults, not a few have been observed in children, such as metaphysical mania, doubting mania, (folie du doute), fear of having committed the unpardonable sin, and recollection mania, which reveal themselves in the child's behavior by an unintelligible inhibition of certain actions only, or of all forms of activity; while others cause the child to perform abnormal actions, as, for example, overscrupulousness which manifests itself in a peculiar, exaggerated exactness and pedantry quite foreign to childish nature, or the fear of getting dirty, as the result of which the child is constantly and incessantly washing itself, particularly its hands (uncontrollable desire to wash).

It appears from the foregoing that the **diagnosis** of the various forms of phobia and uncontrollable ideas may be extremely difficult. The important point is to bear them in mind whenever we are told of any unusual acts of commission or omission in connection with the child, and to try by careful, skilful questioning to gain an insight into the inner (psychic) life of the child.

The **prognosis** in general is not unfavorable. The **treatment** is purely educational.

PATHOLOGIC DREAMING

This is a modern term used to describe phenomena which depend on abnormal imaginative activity. The imaginative faculty is normally much more active in the child than in the adult, as any one can convince himself by observing a child at play. It becomes pathologic only when the things seen and the acts performed in imagination are so vivid as to produce the impression of actual occurrences; the child is under the sway of its daydreams, which determine its actions quite as much as the real things in life. It is a peculiarity of these daydreams that they chiefly relate to the child's own personality and make it appear in a variety of fantastic characters and situations. Since the child fails to distinguish clearly in its mind between dream and reality, a pathologic change is gradually brought about in the "autopsyche." Clinically, the disorder manifests itself, as A. Pick has explained in great detail, in "conspiracies" among schoolboys, the organization of "robber bands," fantastic excursions about the country, etc. The so-called pathologic lie also appears to be produced by this psychic alteration, by virtue of which imagined occurrences are treated as if they were real.

In the experience of teachers there is not so much harm in the kind of imagination that keeps in touch with the reality through the medium of books, dolls and toys, but rather in the imagination which shuts itself off from the outer world and occupies itself chiefly with the child's own personality (Pick). The importance of recognizing this fact is quite obvious from the therapeutic viewpoint. It is not necessary to suppress every imaginative impulse and merely to foster a rational insight into things: but the child must be kept from occupying its imagination exclusively with its own personality by encouraging it to play or by giving it some interesting and rational work to do, such as making collections of various kinds, manual work, drawing and the like, and preventing mental idleness.

DROMOMANIA (PORIOMANIA, FUGUES)

The habit of running away or playing truant is in many cases merely the result of idleness or improper training, without any morbid element.

In other cases, however, punishment both in school and at home is without avail and the habit has a pathologic cause. Epilepsy, which is a frequent cause of similar abnormalities in adults, is rare in children. As a rule a psychasthenia or psychopathic disability is at the bottom of the trouble. Hysteria is said at times to be responsible for similar vagaries (Pick), but we hesitate to adopt his view of these cases.

These attacks of running away, when they rest on a psychasthenic basis, begin in a characteristic manner. The first time, and possibly the two or three succeeding times, the boy (the anomaly is much rarer in girls) is seized by an insurmountable feeling of unrest and is prevented from running away only by fear of punishment, ill treatment or the prospect of hard work. He roams about aimlessly, possessed only with the desire to go as far away as possible, begs his way, if necessary, and after a few days is either picked up in a state of squalor and extreme hunger and sent home again, or returns of his own accord after the psychic storm has abated. Consciousness is perfectly clear during the entire time, memory is intact, and the boy's behavior while he is on his wanderings quite rational. Not infrequently the children resort to lies while on these expeditions or after their return, in order to elicit pity or escape punishment.

In other cases the children run away not from fear, but because they are seized with an intense longing, amounting to "dysphoria," to roam about at their own free will or play with their friends. They often steal money in order to carry out their purpose.

Later on the attacks of dromomania are brought on by the most trivial causes. The dysphoria or ill humor may even come on without any external cause that can be interpreted as a psychologic motive and its advent may betray itself to the attentive observer several days in advance by inattention, irritability and other psychic disturbances. When the dysphoria has reached a certain degree, a nervous discharge takes place and the child runs away. These cases often resemble the epileptic form of dromomania, although careful observation of the child's subsequent life fails to reveal anything that could be interpreted as epilepsy.

During the intervals between the "attacks" many children appear to be quite normal while others exhibit certain defects of character, such as a tendency to lie, cruelty, dishonesty and the like; or they show signs of slender intellectual endowment and are easily led away by bad companions.

In the **diagnosis** of psychasthenic dromomania we must exclude epilepsy on the one hand and mere lack of training on the other. Epilepsy is excluded by the absence of other circumstances pointing to the disease, such as isolated, typical convulsive seizures or, rarely, nocturnal enuresis. The question whether the bad habit is due to lack of training is determined by a study of the boy's environment and particularly his education. As we have already hinted, we do not consider that the hysterical form of dromomania in children has ever been proven.

The **prognosis** depends on the possibility of instituting suitable treatment, which in epileptiform conditions consists in giving bromides, and in the psychasthenic form in educational measures and in guarding the boy from the causes of dysphoria. In order to accomplish anything the boy in practically every case has to be sent to an institution or at least placed under altogether different surroundings.

MASTURBATION (ONANISM*)

It seems justifiable to include masturbation in the group of hereditary neuropathic phenomena, because it is a pathologic condition only when practised to excess and because this is the case practically exclusively in psychopathic individuals.

There is a difference of opinion whether masturbation is very common among children or not, partly because there is no agreement on the definition of what shall be called masturbation in the child. According to Lindner, Hirschsprung and others, it is not rare even among infants and is more frequent in girls than in boys. But according to these two authors any state of voluptuous excitement constitutes masturbation, even when the excitement is brought about not by direct irritation of the genitalia but by sucking movements with the lips, sucking the fingers, the arm, the bedclothes or part of the clothing, or by rubbing or pulling at the ears, picking the nose, scratching the scalp or other similar manipulations.

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* Onanism, or the sin of Onan, is not a synonym of masturbation, according to English usage.

While these vicious habits, which belong among the stereotypias, are no doubt frequently the result of a neuropathic disposition, we shall reserve the diagnosis of masturbation for cases in which there is actual irritation of the genitalia with a resulting orgasm. In cases occurring during the first months or years of life, that is, before there is even a suspicion of sexual feeling—be it ever so abnormally premature and indistinct—we must assume the existence of some organic cause in the genitalia. The most frequent sources of irritation probably are intertriginous processes in the vulva accompanied by itching and, according to a wide-spread belief, oxyuris vermicularis, the worms being sometimes found in the vulva, whither they migrate from the anus. The pleasurable relief from itching, which is at first the only result of scratching and rubbing the vulva and pressing the thighs together, soon engenders a habit that is persisted in on account of the voluptuous sensations which it excites.*

This, however, is not the way children usually begin the bad habit of masturbation. As a rule it is not until some years later, when the child goes to school and is old enough to have a vague, indefinite idea of sexual things, that it first discovers, either by accident or through a playmate, that manipulation of the sexual organs is productive of voluptuous sensations. In itself this is neither morbid nor harmful unless masturbation is indulged in to excess. It has already been stated that the latter occurs almost exclusively in otherwise abnormal children; opinions are divided, however, whether masturbation in such individuals possesses merely the significance of a symptom, or whether it plays a more or less important part in the etiology of the neuropathic constitution by exhausting the nervous system with the repeated excitations terminating in an orgasm. The point is undoubtedly a difficult one to decide, and the arguments offered on either side appear to be based on theoretical considerations rather than on accurate clinical observation. The case reported by Tobler, for example, of a little girl six years old, a member of a healthy family, who kept up excessive masturbation for years without any bad effect on her general health, shows that one is not justified in attributing pallor, a sickly appearance with dark rings around the eyes, headache, an uncertain and capricious disposition, and all sorts of nervous symptoms in a child to a more or less problematical habit of masturbation. We must always assume (in such cases) that there is a congenital weakness of the central nervous system. It cannot of course be altogether denied that in the presence of such a predisposition repeated, violent sexual excitement may add to the mischief; it must be remembered, however, that as a matter of experience—recently confirmed by H. Neumann—the effects of excessive masturbation in small children are comparatively slight. As we are accustomed to regard the nervous system in early childhood as peculiarly sensitive, the fact is worth bearing in mind and should make us scrutinize our cases more critically. Incidentally we may briefly remark that the writers of popular literature on masturbation (Retau and others) fairly revel in exaggerated and most harmful descriptions of the consequences of masturbation.

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* The mere habit of playing with the genitalia, which is observed in small children, can hardly be called masturbation.

It is evident from what has been said that the **diagnosis** of masturbation is quite easy when one has an opportunity of witnessing the act, and in the case of children such opportunities are much more frequent than in the case of older individuals. Descriptions of the act by the parents, particularly if they have themselves indulged in masturbation, must be accepted with reserve.

The **prognosis** depends on the degree of neuropathic constitution present and on the duration of the disorder; it is most favorable in those cases in which sexual sensations have not yet developed.

The **treatment** in the case of young children, before sexual feeling has begun, consists in finding and removing the source of irritation and in preventing the act by means of suitable night clothes, bandages, or some special contrivance such as a pad between the thighs to keep them apart during sleep. In this way the habit is gradually broken up and eradicated.

In older children these mere prohibitive measures are usually insufficient, partly because it is practically impossible to keep the child under constant supervision day and night (in school, in the water-closet, etc.), and partly because even the prevention of the act no longer suffices to eliminate the psychic component (psychic masturbation), which has by that time become more important. In addition to explanation and supervision, dietetic and especially psychic treatment must be instituted with the object not merely of suppressing the perverted imaginative activities, but, by diverting the child's thoughts into other channels and encouraging normal emotions and interests, of gradually displacing the harmful concepts by healthy mental images. In this respect the treatment of masturbation is the same as that of all other psychopathic phenomena (manifestations).

SUICIDE AMONG CHILDREN

A detailed account of such accidents, which must be attributed to some psychic disturbance, is not called for in the present volume.

We may briefly point out, however, that suicide in childhood is more rarely than in adults the result of a true psychosis (especially melancholia), and depends most frequently on an inherited neuropathic constitution. The low resisting power of the mentally unbalanced (déséquilibré) leaves him defenceless against the strain of depression and despondency and the fascination of the thought that death will deliver him from all his troubles. The fact that the instinct of self-preservation, which normally is stronger than almost any other, is overcome—even though it be only temporarily and under the stress of powerful emotion—is in itself a sufficient proof that the suicide is psychasthenic.

HYSTERIA

Aside from a group of special features, which will be sufficiently emphasized in the present chapter, hysteria in childhood does not differ essentially from the same disease in adults. The same difficulty is encountered in defining the limits of this "great neurosis" which separate it from the adjoining territories of epilepsy and neurasthenia. It therefore becomes a duty which requires no further justification to begin by *defining* what we mean by hysterical symptoms, since there is no generally accepted definition that explains the intimate nature of the disease. Investigations by Charcot and his pupils have shown that most of the apparently somatic symptoms of hysteria are really psychic. Möbius expressed this thought in precise terms as follows: "Hysterical symptoms are those which are caused by concepts, including among concepts not only intellectual, but especially emotional states of excitement." According to this view "all hysterical phenomena take the form of suggestions; but some of them, judged by their content are not suggested and represent merely a morbid reaction to emotional stimuli (movements)."

This definition of hysteria has now been accepted by most authors, among them especially L. Bruns, who is an authority on hysteria in childhood, although rejected as too narrow by some, as for example, Binswanger. Charcot himself appears to take a broader view of hysteria and "recognizes in hysteria, in addition to psychic manifestations of disease, other equivalent morbid phenomena which he attributes to nervous or dynamic disturbances" (quoted from Binswanger).

While we do not wish to lose ourselves in a profitless discussion of these controversial questions, we wish, in order to present this subject in as clear a manner as possible, to state at the outset that, in determining what is hysterical and what is not, we adopt the viewpoint of Möbius and refer all "dynamic" disturbances caused by concepts, to the department of neurasthenia, as explained in the chapter devoted to that disease.

By the term concept, let it be repeated once more, we understand not only a clearly defined intellectual mental process but also and chiefly the emotional states.

For the purpose of practical diagnosis it is well to remember that the expression "due to psychic causes" is approximately equivalent to "capable of being simulated." Whatever cannot be effected by exertion of the "will," even after repeated practice or under the influence of intense emotion, is not hysterical. It follows from this definition that there is a *lower age limit* for the occurrence of hysterical disturbances. After the occurrence of typical hysteria before the age of puberty had been recognized as not altogether rare, certain French authors (Chaumier, Ollivier and others) contended that hysterical manifestations may be observed even in infancy. Among these hysterical manifestations there are mentioned emotional symptoms, such as violent maniacal outbursts of rage, syncope, convulsions, conditions resembling meningitis, absence of the conjunctival and pharyngeal reflexes, strabismus, nystagmus, disturbances of respiration, palsies, contractures and the like. Admitting the functional nature of all these phenomena, we must insist that the most important of them, according to our view, belong to neurasthenia and not to hysteria, and that, although Pitres has demonstrated that many children who present symptoms of this kind during infancy later develop hysteria this does not prove the contention. We are justified in regarding the end of the first period of childhood, or say the beginning of the third year of life, as the lowest age-limit for the appearance of hysterical symptoms. It is true that at this early age hysteria is rare and that it gradually increases in frequency after that period. It seems questionable, however, whether this period also is capable of being subdivided into definite stages, especially as it has been maintained that the frequency of the disease undergoes a rapid increase when the child begins to go to school. It certainly cannot be proved by existing statistics because they do not take sufficient account of certain forms of hysteria, which are particularly frequent in childhood and which we shall describe in detail later on. At all events, there are more important factors than school work and increasing age in the etiology of juvenile hysteria. No attempt will be made to give statistics in regard to the frequency of hysteria in the two sexes for the same reasons that none are given for the different periods of life. It may be positively stated, however, that there is a slight preponderance in girls which becomes more marked as puberty approaches and gradually approximates the conditions obtaining among adults.

The **symptomatology** of hysteria is not only variegated but absolutely inexhaustible; "typical" disease pictures are rare in comparison with "atypical" forms. A few characteristic features are common to nearly all cases of hysteria. Familiarity with these features is of great value in the diagnosis because it is a kind of key to the otherwise enigmatical and contradictory observations.

The first thing to be pointed out in this connection is that hysteria in childhood frequently presents itself as a monosymptomatic affection. By this we mean not only the presence of a single symptom but the absence of the somatic stigmata which are so familiar in the hysteria of adults. This behavior, particularly in the case of young hysterical children is not accidental; it is merely the expression of the relatively naïve imagination of the child and is comparable to a similar behavior observed by Krehl among the uneducated agricultural classes in Thüringen and Pommerania. An hysterical child, as Bruns has pointed out, is satisfied with a single symptom and refuses to meet the physician half way when he looks for stigmata. We agree with Strümpell, Bruns, Hellpach and others that the stigmata, such as anæsthesia of a paralyzed arm and the like, usually do not develop until they are sought after, in other words, that they are suggested by the examiner and by the act of examination. "The lay mind of the adult hysterical patient unconsciously reasons that 'a completely paralyzed arm must also be bereft of sensation. If the doctor is looking for sensory disturbances he evidently expects to find them; they belong, so to speak, to the other symptoms which I present;' and accordingly, he actually fails to perceive the pain irritant when it is applied to the arm" (Bruns). The unsophisticated child, on the other hand, is not capable of such a process of reasoning and the paralyzed arm therefore shows no anæsthesia. This dependence of the stigmata on suggestion on the part of the examiner, whether actual or apparently unintentional, is well shown in another group of stigmata, the hysterogenic and hysterofrenic zones and pressure points in paroxysmal forms of hysteria. A convulsive seizure may be produced or inhibited by irritating any part of the body, not only the ovarian region or the testicles, provided the physician has previously predicted the occurrence in a casual remark addressed to other persons in the room. This phenomenon is seen in children old enough to go to school, in whom other stigmata such as, for example, concentric contraction of the visual field, is still difficult to determine and in whom the tests for sensation are uncertain.

The more carefully we examine the more frequently shall we find stigmata, particularly in older children; but the diagnosis must be made without them, because they are, as a matter of fact, frequently absent.

In many cases the picture of the monosymptom itself is of aid in making the diagnosis. It is often characterized by what the French call "massivity"—a gross, exaggerated manifestation of the functional disturbance, which the child, so to speak, thrusts upon the physician's notice. A patient suffering from aphasia at least makes some attempt to speak, while a child with hysterical deaf-mutism does not utter a sound; when asked to speak it fails to make even the faintest movements of the lips. In organic palsy the child is still able to perform a few movements, in hysteria the paralyzed member is absolutely dead and motionless. In a similar manner hysterical pains are characterized by their exaggerated, "insane" intensity, as shown for example when an attempt is made to overcome a contracture by passive movements.

Often the sudden onset of a disease which, when due to an organic lesion, develops more slowly, and its unmistakable origin in some psychic shock (fright, fear) or insignificant bodily injury, suggests the correct diagnosis.

In other cases, the symptoms betray their psychic origin by their contradictory nature with respect to the anatomical conditions. Bruns makes this clear by a number of examples. For example, a paralysis instead of affecting the muscle groups which correspond to its peripheral, spinal or cerebral localization, as in Erb's plexus paralysis or in a cerebral hemiplegia, involves an entire extremity or a segment of the extremity, a hand or a leg, and affects all its movements. Sensory disturbances, instead of exhibiting a segmental character, have a "sleeve-like" distribution which would not be possible in an organic lesion. Everywhere the patient's naïve conceptions of anatomy are revealed.

In the case of young children particularly, the impossibility of the paralysis being due to an organic lesion is often revealed by the fact that only one function of the member is abolished. Babinsky designates this condition paralysie hystérique systématique. The classical example is *astasia-abasia*, in which both walking and standing are impossible although the child is able to move its legs normally and vigorously as it lies in bed.

Add to this the ease with which many hysterical symptoms can be suggested to the child and the frequency with which such symptoms result from imitation of symptoms either in another child or in its own person, which it remembers from some former organic disease—this is so marked as even to attract the attention of laymen—and we have quite a collection of diagnostic points to aid us in recognizing the hysterical nature of a monosymptom.

It is because hysteria has been chiefly studied by neurologists that those forms of the disease which simulate nervous diseases have been known longer and better than any others. But as soon as one has the key which unlocks the mystery of any hysterical symptom, in whatever guise it may present itself, one begins to see a surprising number of conditions which simulate diseases of the respiratory or digestive organs or even surgical diseases. The circulatory apparatus appears to be affected less than any other.

It would take much more space then we here have at our disposal to give a comprehensive and systematic description of the symptomatology of hysteria, and we shall therefore confine ourselves to the most important of the individual phenomena.

Among the *symptoms referable to the nervous system*, the first rank must be accorded to palsies with or without contractures. They manifest themselves in the guise of paraplegia (especially of the legs) or monoplegia of an entire limb or portion of a limb, as for example, one hand, one foot—rarely as hemiplegias or as tetraplegias. As a rule the face is not involved in hysterical hemiplegias. The diagnosis is made by noting that the paralysis, as regards its distribution and the accompanying disturbances (trophic diminution of electric irritability, reactions of degeneration, sensation and the like) presents deviations from the type of a central or peripheral lesion which on anatomic grounds are impossible. In flaccid and in spastic paralysis of the arm the dependent hand is sometimes the seat of œdema, which can be explained on mechanical grounds as a passive œdema and does not necessarily have to be interpreted as a trophic disturbance. Similarly, emaciation of the muscles—always slight—is to be regarded as simple atrophy from disuse.

With regard to the contractures Bruns calls attention to the fact that they are usually characterized: (1) by the extreme degree of muscular contraction; (2) by the intense pain as compared with that which accompanies an organic contraction. They relax during sleep, but return as soon as the patient begins to awake at the examiner's touch. The same behavior is noted when the patient comes out of a deep chloroform anæsthesia. Owing to the intense pain, which is especially apt to be referred to the joints, these conditions have been mistaken for articular neuralgias.

Of other paralytic types astasia-abasia has already received brief mention. Although paralysis disappears entirely during rest in bed, the children are unable to stand or walk when they are taken up and either sink down in a heap or their movements are so ineffective and atactic that normal function is impossible. In this condition also the polymorphous character of hysteria is shown by a wealth of variations in the motor disturbance and its combination with pain in the legs, contractures and the like.

Astasia-abasia, like hysterical palsies in general, usually develops suddenly after a slight injury (a fall on level ground) or a mild febrile disease which has confined the child to bed for a few days; sometimes it comes on after fright. In some cases no cause can be discovered; the astasia-abasia is suddenly noticed in the morning when the child awakes. Young children just old enough to play seem to be chiefly affected.

The hysterical disturbances of speech are both numerous and variegated, presenting most frequently the picture of aphonia and mutism. While mutism, or sudden complete failure to make any attempt at speaking, bears the stamp of hysteria, it is necessary nevertheless, in order to recognize an aphonia as hysterical, that the paralysis of the vocal cord be present only during the act of speaking and disappear during coughing, singing and the like. Such a state of affairs is of course impossible in the presence of an organic lesion. Stammering, stuttering, hesitation and similar articulatory as well as genuine aphasic speech disturbances (Bassenco) may develop singly or combined or in alternation on a hysterical basis. If one bears in mind the general viewpoints enumerated above, the diagnosis is usually not difficult and may be confirmed by the prompt results of suggestive treatment. If suggestive treatment fails or the results are slow, as for example, in the case of hysterical sensory aphasia (speech deafness) in a child seven years old, described by Mann, serious doubts of the diagnosis must arise (Oppenheim).

Blepharospasm in childhood is often hysterical. It usually follows spasm of the eyelids due to the reflex irritation of some inflammatory disease of the eyes and is therefore a kind of hysterical autoimitation or, to use Bruns' expression, the hysterical permanent manifestation of an organic disease. Like all hysterical manifestations produced in this way by autoimitation, blepharospasm is difficult to treat and in that respect, as well as in its origin, resembles the various forms of tic and stereotypia, in which the prognosis is bad. We shall return to this subject later on.

Among motor irritative phenomena contractures have already been mentioned. Choreatic movements are often observed. Many of the relapses of genuine chorea that are of short duration and accompanied by marked muscular hypotonia must be regarded as hysterical, as has already been pointed out in the chapter on chorea. Tremor is not common. The following case reported by Hüssy came under our own observation.

A country boy twelve years of age, vigorous and healthy except for occasional stabbing pains in the chest and back, headache and "shortness of breath." Four weeks ago a tremor began in the hands whenever the boy began to eat, and made it impossible to carry the spoon to the mouth. On examination it is found that he is able to perform complicated movements without any difficulty and writes quite nicely; but when a glass of water is placed in his hand, and he is told to hold it steady or drink out of it, a coarse, violent tremor develops so that he spills the water. The tendon reflexes are exaggerated; pain, taste and temperature sense are normal. A prompt cure was effected by a single painful application of the faradic current, and the boy was able to perform the above-described movements without difficulty. The few cases contained in the literature are cited by Hüssy.

Multiple paramyoclonus (Friedreich), the classification of which in adults is still a matter of dispute, appears to be almost exclusively hysterical in children (Delvart). Only one of the cases so far described later turned out to be a case of multiple sclerosis.

Electrical chorea, to which a separate chapter has been devoted (page 324), may also develop on a hysterical basis. The same is true of rhythmical chorea and facial spasms, and the various forms of tic, which are described at length in another place.

The convulsive seizures form an important group of hysterical manifestations. They may take the form of chorea magna as described by Charcot and his school, but usually occur in older children, and in that case are practically identical with grand mal hystérique, the diagnosis of which presents no difficulties. When the attacks resemble epileptiform seizures, however, the diagnosis is not so easy. They may be unilateral, confined to a single extremity, or universal; they may be ushered in by a kind of aura and may be followed by peculiar confusional states and the like, thereby suggesting a genuine or a cortical epilepsy if the physician has to depend solely on a laymen's description.

But if the physician has the opportunity of observing the attack himself, which is often possible at the first visit because the attacks are readily brought on by suggestion, he is soon convinced that the alleged unconsciousness, which the parents usually infer from the fact that the child does not answer, in reality does not exist, that the pupils react, and that the character of the muscular contractions and convulsive attitudes is much more complicated and theatrical than in epilepsy. Moreover, the sphincters practically never relax during hysterical convulsions, so that there is no involuntary discharge of urine or feces, nor does the child bite its tongue or do itself any other injury in falling. The presence of the three last-named symptoms is much more in favor of epilepsy. Cases of hystero-epilepsy, by which is meant an intermediate form between the two neuroses or a simple association of the two neuroses in the same individual, have, so far as I know, never been observed in children, although their occurrence is quite as conceivable as in adults. It is worth noting that in many cases the daily return of the attack at exactly the same hour at once awakens a suspicion of hysteria. For the rest, the diagnosis is determined by the general viewpoints which have already been described.

The delirious states which frequently accompany or alternate with the hysterical convulsive seizure may occur independently in children. Henoch in his conferences describes cases of this kind with sudden and usually terrifying delirium, with violent ravings and loud outcries, which appear to be produced by terrifying hallucinations or visions. The same observation has been made by Bruns, Eulenburg and a number of French authors.

The pronounced theatrical element which characterizes these forms of delirium as well as the stuporous states and the hysterical forms of somnambulism usually renders the diagnosis clear at once. In the main these severe forms of hysteria in older children, not excepting the occasional prominence of sexual and erotic concepts, so closely resemble the manifestations of hysteria in the adult that a separate description appears to be superfluous in this place.

Among disturbances of the special senses, deafness and blindness of one or both eyes are the most frequent in childhood, but they are absolutely rare manifestations of hysteria at that period of life. For one who is acquainted with these phenomena in the adult no special description is necessary.

In connection with the forms of hysteria which imitate nervous diseases brief mention may be made of a few manifestations simulating surgical diseases of the bones, joints, muscles, tendons, etc. Contractures of the muscles of the front and back of the neck, when unilateral, may simulate torticollis, and when bilateral, cervical spondylitis. Elevation of one shoulder, rigidity or kyphoscoliotic curvature of the vertebral column, in which attempts at reduction are accompanied by fixation of one hip-joint as in beginning coxitis, and many other symptom-complexes of the same kind may be produced by hysterical muscular spasms. The diagnosis in practically every case can be made on general grounds, particularly on the sudden onset after some insignificant physical injury or psychic insult, the "massivity" of the pains, and the absence of any cause for a corresponding organic disease.

The hysterical pictures which simulate diseases of the respiratory organs as a rule develop from corresponding organic affections by auto-imitation, as above briefly described, or as a permanent manifestation of the antecedent disease. Although they have only recently begun to be properly appreciated (Thiemich), and we have only a few isolated reports from earlier times, they are well worthy of careful attention on the part of the physician. For example, if a child has ever suffered from tachypnœa and dyspnœa as the result of extensive bronchitis, it may later, whenever it has the slightest catarrh, exhibit the same symptoms either as psychic phenomena or through an hysterical reproduction of its former diseased state. In an entirely similar manner a spasmodic cough, possibly with violent stridor, an asthmatic attack and the like may be reproduced hysterically. In order to recognize this change in the condition of affairs, which is particularly difficult for the attending physician who has seen and treated the patient during the primary organic disease, a painstaking objective examination must be made, bearing in mind the general diagnostic features of hysteria. The importance of making a correct diagnosis from the therapeutic standpoint will be referred to later.

Hysteria in children is not infrequently localized in the *digestive tract* and associated organs. Anorexia, vomiting, cardiospasm, diarrhœa, constipation, meteorism, incontinence of feces, prolapse of the anus and the like may be due to hysteria. The anorexia, which is probably primarily produced by some organic disturbance, may in severe cases (Soltmann, Kissel, Bruns and others) go on to an alarming state of inanition threatening the child's life. In many cases the refusal to take food can be overcome by simply placing a tray in front of the child instead of trying to force it to eat. When the child finds it is not under observation it will satisfy its hunger of its own accord. In other cases gavage will not only prevent the extreme effects of starvation but also affect a prompt cure of the anorexia through suggestion. Hysterical *vomiting* is usually recognized at once by the ease with which the act is performed, just like a volitional act. Merely awakening the child in time for school or a harsh word may suffice to bring on the vomiting. The prompt result which follows a single introduction of the stomach tube for diagnostic purposes confirms the diagnosis of hysteria. An instructive case of cardiospasm in a child two years old, which was also promptly cured by passing a tube, was observed in the Breslau Children's Hospital (Freund). Psychic *diarrhœa,* that is to say, evacuation of feces, normal except for their frequency, sometimes persists a long time after the subsidence of an organic intestinal disease. It resists all astringent remedies but yields at once to faradization of the abdomen, a procedure which has been soberly recommended as a treatment for diarrhœa although it cannot act in any other way than by suggestion. Constipation may develop in the same way, or rather may become a fixed habit from psychic causes, even aside from those cases in which the child has simply forgotten how to evacuate its bowels spontaneously.

It must be pointed out, however, that *constipation,* as well as anorrexia and vomiting for that matter, may be caused by an imperative idea or, in other words, may be due to psychic but not hysterical causes. Thus, the fear of being poisoned is often present in anorexia. In two cases of retention of feces, observed by Neumann in young boys, the cause was found to be disgust and loathing of the act of defection. "It is nasty to do Aa" said a little patient aged three, who would never tell the nurse when his bowels needed to be moved and would not evacuate when told to do so except at night when the inhibitory concept was in subjection.

Hysterical *meteorism* of a pathologic degree results either from swallowing air—in which case there is always a suspicion of simulation—or from nervous tonic spasm of the diaphragm, and is particularly apt to be mistaken for peritonitis when it is associated with marked sensitiveness of the abdomen. Cases due to spasm of the diaphragm (pseudotympanite nerveuse ou ventre en accordéon, Bernheim), may be recognized by the fact that the abdomen collapses under chloroform anæthesia, without the escape of gas from the mouth or anus as when the condition is due to swallowing air.

The recurrence of prolapse of the anus as the result of some fixed idea, which in children is usually quite transparent, has also been observed in a number of well-authenticated cases in children about the age of three.

Finally we have two disturbances in the genito-urinary apparatus that are frequently hysterical: *Pollakiuria,* that is, the evacuation of urine at abnormally frequent intervals, and diurnal and nocturnal *enuresis.* As both symptoms may be due to a number of organic diseases affecting the nervous system, metabolism and the genito-urinary system, these causes must first be excluded by a careful physical and urinary examination. But even after the functional nature of the disturbance has been established by exclusion, the diagnosis of hysteria is not justifiable without further proof.

This subject, which has been extensively studied in the past few years, is sufficiently important to justify a more detailed discussion. While French authors (Janet, Guinon and others) had long recognized and insisted upon a psychic etiology in enuresis, the majority of German authorities (see for example the text books of Henoch and Baginsky) regarded the condition as a local neurosis of the bladder, and the only controversial point was the mechanism of the disturbances or its mode of onset. Czerny's school was the first in Germany to insist upon the psychogenic origin of enuresis as well as of pollakiuria, and the analogous disturbance of incontinence of feces, which they explain as hysterical symptoms. This is undoubtedly true in a certain proportion of cases of enuresis and is proven by the occasional sudden occurrence of enuresis or pollakiuria in children who had not been subject to it in infancy. In these cases there is evidently a definite cause, as for example, in one of Reinach's, as is shown by the prompt and permanent result which followed antihysterical treatment (see page 378). In the same category belong all the cases of psychic infection, in children who had previously been free from enuresis, by other children in the same class or boarding-school who are affected with the disease, and the promptness with which cases of this kind due to unconscious imitation recover, is a strong argument in favor of the hysterical nature of the disturbance. Pfister, on the other hand, advanced a different view in an important paper on the subject in which he cited a number of cases of enuresis. He has, in addition to the cases due to organic causes, a group in which the nocturnal enuresis represents an abortive or unnoticed attack of epilepsy. This group is characterized by the fact that the enuresis occurs "quite sporadically, possibly for a short period every night in succession, with intervals of irregular duration, from several days to many weeks or even longer when the child is quite free from the disturbance." On careful examination other signs of nocturnal epileptic explosions are always discovered in adults and usually also in children (he refers to children of at least five years of age or older). Among these signs are biting the tongue, falling out of bed, sometimes pronounced lassitude on waking up in the morning, and the like. This form of enuresis with a suggestion of epilepsy is also mentioned by Binswanger and others. In Pfister's second group the enuresis begins in infancy and the children can never be trained to cleanly habits: or it begins some time between the third and sixth or seventh year of life with frequent and usually nightly evacuations at different hours of the night. In addition to the nocturnal evacuation, for which it is frequently impossible to discover any special cause (possibly dreams?), there is usually also some diurnal enuresis which, however, is much more rare and is brought on only by fright or inattention when the child is busily engaged in play. This variety of enuresis, which is extremely variable and is often associated with pollakiuria and other functional disturbances of urination or even incontinence of feces, and which obstinately resists all educational therapeutic measures, is regarded by Pfister as a sign of functional degeneration and an hereditary neuropathic stigma. The arguments which he advances in support of his theory, the details of which are extremely ingenious, are too lengthy to be reproduced in this place; but it seems desirable, in order to throw light on the pathogenesis of enuresis and related disturbances, to mention at least his main proposition, the general neuropathic etiology and significance of the affection. If this is borne in mind, all other causal factors that have been mentioned, such as adenoid vegetations, the drinking of large quantities of fluid in the evening, cold beds and consequent chilling of the abdomen, slight affections of the bladder (Rey) or changes in the composition of the urine (Lawrence and others) assume at most an accidental significance. While it seems to us that Pfister in his character of psychiatrist has exaggerated the frequency of the severe, degenerative cases, his writings nevertheless represent a distinct advance in therapeutics. He has shown us the necessity of differentiating our cases more accurately. Whenever there is any reason to suspect epilepsy, the corresponding treatment and supervision of the patient must be begun at once. On the other hand, in functional disturbance of the bladder in which epilepsy can be positively excluded antihysterical treatment must be instituted as rapidly and energetically as possible in a manner to be presently described, and in this way prompt and permanent results will be obtained in a great many cases. Nevertheless one must not be surprised if the treatment fails at least partially in a great many cases. These will be put down as belonging to the degenerative form, in which general hygienic and educational measures must be tried, although time and the gradual bodily and mental development of the child will in the end prove the most potent factors in bringing about a cure. Accordingly we may dispense with a mere enumeration of all the various infallible remedies which have been recommended without the slightest attempt at discrimination. There is no specific remedy. All methods of treatment act either by suggestion or not at all, including epidural injections (Kapsammer), which are so greatly lauded by Cathelin. Even the results of hypnotic suggestion are neither better nor worse than the results obtained by any other form of psychic treatment (Hacklander).

The **etiology** of hysterical manifestations, the most important of which we have just described, might be given more briefly if we were better acquainted with the nature of hysteria, which is but imperfectly characterized by such expressions as limitation (narrowing) of consciousness, increased susceptibility to suggestion, an abnormal tendency to exhibit somatic reactions to psychic influence and the like.

The most important etiologic factor, heredity, in itself gives us no information in regard to the disturbance that is going on in the mechanism of the child's psychic life. The causal significance of environment and education is rather more intelligible, but here the question at once presents itself whether these injurious influences really produce the foundation of the malady, the hysterical alteration of the nervous system, or whether they merely act as exciting causes—as "agents provocateurs"—and bring the disease to the surface. The influence of somatic diseases also in this respect is by no means clear.

The fact that hysterical symptoms may appear as the result of inadequate causes forces upon us the theory of a latent condition. A correct understanding of this latent condition or at least of its psychopathologic signs, would be of the greatest value; but that is as yet beyond our reach.

Numerous attempts to explain this condition have been made. The French, especially San Philippe and others, have described a hystérie latente and a hystérie naissante, but the conditions to which they refer are in the main neuropathic or genuine neurasthenic changes in character, such as we frequently observe as forerunners or concomitants of hysteria in children. Unfortunately these traits are often missed in the purest cases of hysteria.

On the other hand it seems quite justifiable to speak of an hysterical character, which consists in exaggeration of the emotional life, a lively imagination coupled with an unchildlike interest in the process and conditions of the body, egotism and an instinctive desire to appear important, to attract the attention and, if possible, excite the admiration of other persons. These traits of character help to explain the fantastic confabulations of hysterical children, and especially their mania to utilize their morbid symptoms in making trouble for persons whom they dislike as, for example, when they develop an hysterical palsy after they have had their ears boxed by a strict and exacting teacher. The hysterical character may develop in the absence of any lack of conscious or unconscious education, and in these cases it must be assumed that the predisposition is unusually pronounced. As a rule, a vicious environment is at least partly responsible for its development. In many cases the hysteria can be shown to be due to direct imitation of diseases of the parents; but the most important cause is the wrongly-directed, usually too indulgent and always capricious education of the child by hysterical or neuropathic parents. This point requires no further elucidation. In other cases external insults such as fright or fear are operative causes. Thus we once observed a state of deaf-mutism lasting several weeks in a little boy of three who had been frightened by the sudden apparition of a cat on a dark stairway. Bruns, for example, observed unilateral convulsions resembling night terror coming on in the morning as the child was called to get up and go to school, and for which the child's fear of a strict teacher was responsible. The same authors cite examples of hysteria in children whose fathers were drunkards and abused their wife and children when intoxicated, and points out that in these cases the desire to escape the unhappy home life by being admitted to the hospital is also a factor in the etiology.

Autoimitation of organic diseases, which has been referred to repeatedly in this section, is absolutely fostered by unintelligent, overindulgent parents who let the child have its way while it is ill and gratify its most foolish wish, exhaust themselves in expressions of love and pity, and do everything to make sickness a condition to be greatly desired by the child. An hysterical disease in a child surrounded by such an environment has many points in common with conscious simulation or may, in fact, be nothing but simulation. As physicians, however, we must remember that the simulation itself is a pathologic trait of character. A child that is psychically normal does not simulate disease.

The degree of intelligence necessary for the simulation of hysterical symptoms is by no means high, while it is true that many hysterical children are more intelligent and vivacious than average children of their own age, and accustomed by their constant association with adults to observe and form judgments in a way not usual with children—in short, are what is generally known as old-fashioned. Hysterical disturbances nevertheless occur in children of very slender mental gifts and in imbeciles. An interesting fact in this connection which was brought out by Bruns and which we have also been able to confirm is that the grossest, most "massive" forms of hysteria occur chiefly in country children who have a very limited mental horizon.

There is but little to add about the **diagnosis** of hysteria. The frequency with which hysterical traits are associated with organic diseases is a warning to use the greatest care in the objective examination of the patient. Thus in many forms of brain tumor (glioma with periodical bleeding, intraventricular cysticerci and the like) the clinical picture may be very deceptive and closely resemble that of hysteria. Failure to recognize an hysterical symptom as such and accordingly to apply the proper treatment greatly diminishes the chance of recovery by confirming the parents and attendants in their belief that the child is suffering from a grave disease. For the physician himself the mistake is serious only when another physician or even a quack is called in and by recognizing the true state of affairs rapidly brings about a cure. Conversely, parents will never forgive a physician if he fails to recognize an organic lesion and calls it hysteria, because in the lay mind this word always has in it an offensive element of simulation or of over-anxiety that need not be taken seriously.

The **prognosis** of hysteria in children is in the main *favorable*, not only as regards the individual manifestation, which is often quite easy to cure, but also the psychic constitutional anomaly as a whole. This important distinction between hysteria in the adult and the same disease in children was first established by Bruns, who kept his patients under continual observation after their recovery. He states distinctly, however, that complete permanent recovery is effected only when the first hysterical symptom is recognized as early as possible and removed by appropriate treatment or as he says, extirpated. As the result of numerous unsuccessful therapeutic experiments the first symptom becomes inveterate or the psychic anomaly so firmly rooted that it never disappears altogether. Even if the individual symptom is ultimately removed, other symptoms sooner or later make their appearance and betray the fact that the hysterical change of character is permanent. The prognosis is undoubtedly most favorable in the hysterias which are produced by imitation or psychic contact infection. These forms are sometimes epidemic in schools as, for example, hysterical chorea (Holwede), hysterical tremor (Demmer). The children who are attacked secondarily always recover rapidly in such cases.

Treatment.—It follows from what has been said that the treatment of hysteria in childhood is purely psychic. The physician is rarely able to prevent the development of the constitutional disposition, which is due to heredity; but he can often see to it that the child is separated from its hysterical mother, either permanently or at least during her attacks, or that the governess, if she has hysteria, is dismissed. But above all he must, by refraining from every kind of treatment that is not absolutely necessary, guard against fostering hypochondriac introspection and self-indulgence which lead to hysterical manifestations. He must also exercise an advisory control of the child's education.

The treatment of pronounced hysterical symptoms offers to the physician a wide field for the exercise of whatever he may possess of ingenuity and sagacity, as well as tact and sympathy. We strongly recommend Bruns' comprehensive and stimulating writings on this subject and are content in this place to indicate merely the proper lines which must be followed in the treatment. In young children, who are in the main credulous and used to obeying, the treatment may be purely or, as Strümpell says, undisguisedly psychical. Thus, in a case of astasia-abasia the child may be simply set on its feet with the brief injunction "now walk" or the like. But in most cases the physician finds himself compelled to mask the psychic effect of his treatment by employing physical methods. The most effective are those which have a strong suggestive action, such as pain or, on the other hand, whatever must appear mysterious and wonderful to the child and thereby render it susceptible to suggestion.

We shall adopt Bruns' suggestion and subdivide methods of treatment employed in the hysteria of children into *two groups;* one method consists in *taking the child by surprise,* the other may be called the *method of intentional neglect,* or ignoring the child. Each of these methods has its special indications. The first is particularly effective in all cases of paralysis and contractures, in aphonia, deaf-mutism, etc.; the other is more appropriate for irritative conditions, especially those which occur paroxysmally as spasms, delirium, somnambulism and the like.

When the plan of *"taking the child unawares"* is to be employed, the hysterical symptom is attacked directly, either undisguisedly as we have just shown in the example of astasia-abasia, or under the guise of some physical method of treatment (with the faradic brush, the cold douche, by forcible extension of a contracted joint) and if possible removed at a single sitting. If this proves successful, the result is usually permanent; if not, or if the success is only partial, "the child may recover from its first surprise and astonishment at the doctor's actions and the rapid results affected," and what remains of the symptom as a rule becomes firmly rooted and can no longer be influenced by this method of treatment.

The second method, that of *intentional neglect,* which, as has been stated, is chiefly to be recommended in paroxysmal forms of hysteria, must then be resorted to. This method obviously requires so much more time, so much intelligent coöperation on the part of the parents, and so much judgment to determine whether and to what extent it is necessary to take any notice of the morbid symptom, that it can rarely be carried out successfully at home and usually necessitates removing the child to an institution. The object of the method is to convince the child of the harmlessness of its disease by paying no attention to its spasms and other symptoms. It is the direct opposite to what the child is accustomed to receive at home, surrounded by the overanxious care of its excited and exciting parents. "If the symptoms no longer attract attention, says Bruns, they gradually die of ennui; the child forgets them, so to speak, altogether." Whether the symptoms are to be ignored altogether or intentional neglect is to be combined with secondary methods, such as hydrotherapeutic procedures, douches, wet packs and the like or by faradization, must be determined in the individual case by the physician's practical experience.

Isolation—not solitary confinement but removal from the accustomed surroundings—is another important procedure. Isolation is often necessary not only for the proper carrying out of other methods of treatment but because it has a curative effect in itself by feeding the child's imagination with new, healthy impressions and neutralizing the injurious influence of the environment in which the hysteria developed. The greatest resistance is, as a rule, encountered on the part of the parents when the physician proposes isolation. Incapable of realizing that they have unconsciously injured their child, they are convinced that it will only be made worse by homesickness and the grief of isolation, and are therefor ready to make any sacrifice rather than consent to separation. As the mere dread of a prospective isolation often has a wholesome suggestive influence on the child, it is not wise to propose isolation as the only means of salvation; but the physician should recommend it as early as possible before he has destroyed all chances of recovery by the failure of innumerable therapeutic experiments, not only on his own account but also on account of every other physician who may be called in after him and may be under the additional disadvantage of being a stranger to the child. Once the physician has decided to insist upon isolation he should, as a rule, avoid entering into a prolonged and fruitless discussion of the advantages of the plan with the parents, but, as Bruns points out, simply take his stand on his experience as a physician.

Aside from the above-mentioned advantages, treatment in an institution has the additional advantage that all unpleasant methods of treatment, hydrotherapeutic procedures and painful faradization, which are often extremely useful, can be carried out much more easily than in the presence of the excited and anxious parents. In regard to these measures, however, we must insist that cruelty is neither necessary nor justifiable, and that the pain-inflicting treatment must never lay aside its mask and never be allowed to appear to the child as a simple punishment. If the child sees in the physician its enemy and tormentor, all suggestive influence is lost.

We may illustrate this by a personal observation. A boy about ten years of age was admitted to the hospital on account of severe hysterical spasms for which he had been repeatedly treated without success, and in the hope of showing good and rapid results the boy was subjected to painful faradization after every attack. Although the boy was very much afraid of the treatment, the spasms refused to yield, becoming worse rather than better, and the boy was taken away about ten days later no better than when he was admitted, after treating the father to a striking attack on the occasion of his first visit. On leaving the clinic the father, who believed the condition was epilepsy and indignantly refused to accept the diagnosis of hysteria which had been written on the boy's card, went to a neurologist in this city who accepted our diagnosis and proposed that the boy be admitted to his private clinic. The father could not bring himself to consent to this at once and took the boy home for another trial, promising to bring him to the clinic if the spasms should return. From this time on the boy never had another attack. He was cured by the fear of being sent to another hospital. This is all we could learn in regard to the patient, whom for obvious reasons we never saw again.

We do not of course wish to deny that treatment sometimes fails even in a hospital. Many of these patients recover through some "miraculous cure," by a quack, or as the result of some fortunate accident as in the case just described. In others the symptoms subside gradually as in the hysteria of adults. Of the prognosis in such cases of course nothing more can be said. Under certain circumstances it may be justifiable to employ hypnotic treatment as a last resort.

We are as much in the dark on this point as in regard to the nature of hypnosis in general. While the school of Nancy (Bernheim and his followers) regard hypnosis as a perfectly harmless procedure when carried out in a rational, scientific manner, and recommend it as a pedagogic measure in the moral education of the child (Berillon), the school of the Salpétrière (Charcot, Gilles de la Tourette and others) adopt the opposite view. They consider the hypnotic sleep an hysterical condition and believe that it may be followed by results that are much more serious than the disease for the cure of which the hypnosis was employed. At all events the hypnotizer must be an expert in his line, and the method must be limited to cases of the greatest gravity and then only employed as a last resort. In Germany hypnotism has rarely been employed in the treatment of infantile hysteria by responsible and serious physicians, and it is not likely that it will be more extensively employed in the future, especially as its popularity appears to be waning.

PSYCHOSES

The most important of the psychoses in childhood are those which are associated with defective intelligence—so called *defect psychoses*. Among these the active congenital conditions which are included under the general term of "*imbecility*" are much more frequent than acquired dementia.

Congenital defect psychoses are peculiar inasmuch as they usually represent the symptoms of an organic brain lesion and are therefore permanent, irreparable anomalies.

An exception to this is found in one group of congenital defect psychoses which is produced by *disturbances of the function of the thyroid gland—cretinism* and *myxidiocy* in its various forms, which are susceptible to organotherapeutic treatment and therefore not caused by an anatomic brain lesion. As these conditions, including *mongolism* which from the viewpoint of differential diagnosis belongs in the same group, have been described in connection with diseases of the thyroid gland, all that remains to be added here is a short description of one symptom, namely, the impairment of intellect, without regard as to whether it is associated with other symptoms or constitutes the entire clinical picture.

We distinguish three degrees according to the severity of the disturbance; *idiocy, imbecility* in the narrower sense, and *feeble-mindedness* (debility). The lines of division are by no means sharp. Absence of attention is said to be characteristic of the idiot, while the ability to follow quite a complicated train of thought with a preponderance of ethical deficiency is usually said to be the distinguishing mark of the feeble-minded.

There is never any difficulty in recognizing idiocy, at least after the first few months of life. The absence of reactions to impressions from the environment that indicate the awakening of the mind is proof enough.

In the case of imbeciles greater difficulty is experienced because, in the beginning at least, the absence of reactions is not complete and there is simply delayed psychic development. While it is possible in an imbecile infant of about six months to attract the attention by making a noise, by holding up bright objects and other similar methods, the attention cannot be fixed because the new impression finds no acquired impressions with which to associate itself and accordingly fails to arouse any interest. Hence, these children, although their muscles are well developed, do not hold their heads up nor grasp things nor laugh, and are late in learning the coördinated movements of sitting, standing and walking. Even with the greatest care it is often impossible to train them to cleanliness until the third year or later.

Another very important early symptom, which is also explained by the absence of attention, is the diminution of the pain sense. The sense of taste is often so greatly reduced that the children will swallow sour and bitter solutions without making a face. In itself, however, the disturbance of taste is not as conclusive a proof of imbecility as is hypalgesia, because it also occurs in children who are not imbeciles but are suffering from chronic rachitic disturbance (rachitis) during the first two years of life. While the positive demonstration of hypalgesia and hypogeusia are exceedingly valuable, the absence of these anomalies is by no means proof that the child is psychically intact. Moreover, there is no constant relation between the disturbances of pain and taste and the degree of the mental impairment in other respects.

As the child becomes older the next most important criterion, the development of the faculty of speech, becomes manifest.

While a child with normal mentality begins to speak at the age of about eighteen months, unless its development has been greatly delayed by disease, an imbecile often does not begin until it has reached the age of three or four years, and its progress is very much slower than that of a healthy child. Again, however, there is no strict parallelism between the development of the speech faculty and mental impairment. Some children begin to speak at the normal time and make fairly good progress; while, conversely, in children with mental impairment so slight that it is overlooked for a long time or possibly overestimated, the disturbance of speech appears to be merely an isolated aphasic disturbance (word-deafness). In cases of this kind the child understands what is said very well for some time; but while in the normal child this stage is shortly followed by the actual ability to speak, that power is developed very late and very slowly and the child apparently never learns to speak properly. The same disproportion as between speech and intellect in general is often found between the various other intellectual faculties. Gross defects in one or more departments of intellectual activity may be associated with normal or even unusually good development in others; for example, a feeble-minded child may be very good at arithmetic because it possesses an unusual and overdeveloped memory for numbers; another may be musical; a third an unusually clever actor and entertain the other children by the performance of all kinds of tricks. Another type that is quite common in institutions for the feeble-minded is seen in those children who learn to write and read without any difficulty but have no idea whatever of numbers and, after years of schooling, are barely able to do a simple sum in arithmetic involving no more than the addition of numbers up to ten, or even to count the fingers.

The disturbance evidently depends on absence of memory and resulting inability to associate memory pictures with the simplest mental impressions. Pronounced imbeciles are unable to distinguish between their nearest relatives and strangers; they do not know their own clothes and cannot find their way about, etc. In the milder grades the child is able to perform these simple acts, but when it is tested with objects or pictures, it is found to be unable to recognize or designate objects of daily use and toys. In so-called "word-deaf" imbeciles the examination must be confined to naming the desired object and asking the child to point it out, if the error of overestimating the degree of mental deficiency is to be avoided. Great inaccuracy of observation and confusion of mental images are frequently observed. While the child is able to distinguish a dog from a goose in a picture book, it will confound a dog with a cat or a goat or a goose with a duck or a stork. The color sense is also uncertain and late in developing.

Special conceptions, as for example, above, below, larger, longer, smaller, shorter and the like are usually defective or entirely absent.

It is the same with the causal connection of various things, for example, if we ask such questions as proposed by Ziehen—"why do we heat the house in winter?" or "why have I brought an umbrella with me?" an imbecile child is rarely able to give a suitable answer.

Higher abstract ideas such as duty, ownership, envy, gratitude, good, bad, are always absent in imbecile children. The emotional life also is usually characterized by the poverty and temporary character of the emotions.

This impairment of the emotional life is particularly characteristic of feeble-minded children, in whom the impairment of intellect may be slight or limited to only a few mental functions. The ethical impairment is the most conspicuous trait of feeble-minded children. Friendship, attachment, gratitude, respect, sense of duty, love of truth and the like are very feebly developed and usually, after a few attempts at correction which make very little impression, are replaced by the opposite evil impulses (moral insanity).

Intellectual impairment is less marked in feeble-minded children, so that they are able to carry on quite complicated trains of thought and perform quite intricate acts, crafty intrigues and lying excuses. As a rule it is not recognized until the child begins to go to school when it is found to be unable to keep up with other normal children of the same age and falls behind in spite of additional help and the most careful teaching. In the milder grades the child while in the lower classes appears to be normal, but fails to make good when it reaches the middle or upper classes where, in addition to exercises which chiefly tax the memory, work demanding higher intellectual reasoning powers is required. These cases which are recognized late, when the child has almost reached the age of puberty, must not be mistaken for acquired dementia (hebephrenia and the like), which often exhibits a progressive character.

Medical treatment is required only in those rare cases of imbecility in which the mental deficiency is a symptom of disease of the thyroid gland or possibly of some organic brain disease as, for example, brain syphilis, that is not altogether incurable. If these conditions cannot be excluded, an attempt should be made to treat the primary condition.

For the rest, the treatment of imbecility belongs to the pedagogue, whose duty it is to develop what there remains of mentality by judicious guidance and suitable exercises. This field has been developed beyond all expectation in the last few years and already possess an almost unlimited literature. The limits of this work forbid even a fragmentary exposition of the methods that have been worked out and the results that have been achieved in this field by the coöperation of physicians, psychologists and pedagogues.

Acquired dementia occurs among children in various forms. Two of these forms, paralytic and epileptic dementia, have been mentioned in other portions of this book. Dementia occurring with focal disease of the brain and hebephrenia (dementia præcox) do not require special description. The former differs in that it is not congenital, and does not appear before the acquisition of some mental state of possession; while hebephrenia is a disease of puberty or, in exceptional cases, has its inception shortly before that period. The reader is referred for this subject to the text books on psychiatry.

Among the psychoses without defective intelligence we have mania and melancholia, both as simple diseases occurring only once and as periodical or circular forms. They are very rare before the beginning of puberty.

Acute *hallucinosis* or amentia, chronic paranoia, stuporous conditions ("twilight conditions") on an epileptic or hysterical basis, and the forms of delirium in severe febrile diseases (such as the delirium of incubation, or collapse) inanition and intoxications (called by Ziehen concomitant delirium) are practically the same in children as in adults and therefore do not call for any special description. Moreover, like all functional psychoses, these conditions are rare in children.

Uncontrollable ideas and *actions* have already been discussed in the chapter on hereditary neuropathia. In the same connection psychopathic deficiency was also mentioned. The psychotic conditions which develop as the result of these anomalies and which exhibit a special character are sometimes observed as early as the later periods of childhood and merit attention on account of the frequent collisions that they produce in families as well as in civil life.

DISEASES OF THE MENINGES

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THE anatomy of the meninges in childhood does not differ materially from that in adults.

Of the pathological changes the acute and subacute, due to bacterial inflammations, chiefly interest us; after these come the chronic forms, serous meningitis as a consequence of angioneurotic disturbances (first observed by Quincke), and meningeal tumors, discussed elsewhere.

With the exception of headache, which is a consistent though dubious symptom in young children, the *clinical symptoms* upon which we base a diagnosis of meningeal disease are secondary, due to pressure upon, infiltration or circulatory disturbance of, the underlying tissues. These conditions evoke numerous manifestations of irritation and paralyses, the manifold combinations of which present the various clinical pictures which will be discussed.

There is no pathognomonic symptom, which by its absence, precludes the diagnosis of meningitis. However, there appear in the confusingly varied field of vision some symptoms which, on account of their frequency and relative importance, deserve to be advanced as *cardinal symptoms,* prior to a discussion of the various froms of meningitis *classified etiologically and pathologically.*

The first symptom is *headache,* a symptom hardly ever wanting in meningitis, at any rate in a child more than a year old. Comment upon the significance of this symptom is unnecessary.

The second symptom is *disturbance of the sensorium,* or, as usually learned, psychical disturbance. Beginning with change of disposition, as for example disinclination to play, peevishness, and repugnance to occurrences and impressions formerly disregarded (as eating for instance); proceeding to drowsiness and weariness without actually sound sleep, then to apathy which can be overcome only with difficulty, finally to deep reactionless coma, this disturbance runs a most varied, and as must be emphasized, a most enigmatical course. But, it is important to keep in mind that it is never absent when there is continuous observation.

As a third symptom we may mention *fever.* We must, however, except hæmorrhagic pachymeningitis and Quincke's serous meningitis;

and we must emphasize the fact that fever may be entirely absent throughout limited periods of observation and for long periods it may be slight. It is unnecessary to dilate upon the ambiguity of this symptom; however it may not be superfluous to recall how frequently in any single case a most careful observation fails to explain fever of days or of even weeks duration in children.

Frequently, though not invariably, inflammatory processes in the meninges cause increased *intracranial pressure*, with more or less constant symptoms. These are vomiting (cerebral), disturbances of the pupillary reaction, unevenness and irregularity of the pulse, brachycardia at the beginning and tachycardia (explained by paralysis of the vagus nerve) toward the end of the disease, hypertony of the muscles, etc. A special form of hypertony occurs almost invariably when the meningeal process is localized in the occiput, that is *rigidity of the neck* (opisthotonos). This symptom appears in pronounced cases as a painful contraction of the cervical muscles, which draws the head backward so that it bores into the pillow; in milder cases there is opposition (offered reflexly as the result of pain) to passive forward, less to lateral movement or rotation of the head. Hypertony can be detected in the milder cases also by Kernig's phenomenon—(when the patient sits upright the knee, on account of spasm of the flexors, cannot be straightened). It is noteworthy, particularly when the spinal meninges are considerably involved, that pronounced spinal rigidity may ensue and likewise induce Kernig's symptom. Cervical and spinal rigidity are very valuable symptoms of meningeal affection when the other causes (spondylitis, muscular rheumatism, enlarged glands associated especially with nasopharyngeal disorders (Pfeiffer's glandular fever), and hysteria can be excluded. However, in very many cases it does not occur. It it evident that increased intracranial pressure is most trustworthy as a guide in the diagnosis of meningitis when it is directly demonstrable and not deduced from other symptoms, which are themselves capable of various interpretations. This is possible in infants with sufficiently large anterior fontanelles. In fact the daily increasing bulging and tension of the fontanelle is one of the most reliable signs of meningitis in early life, and its value is still further enhanced by the fact that at this time it is difficult to establish other symptoms referred to or rely upon them on account of their ambiguity. Increased tension of the fontanelle, however, is indicative of meningitis only when the child is quiet and if moreover other causes, pneumonia, etc., can be excluded. We have seen a rigid, pulseless fontanelle notwithstanding severe depletion of fluids following diarrhœa in a case of sinus thrombosis, in a septic infant, without meningitis.

If, in conclusion, we remark that under certain conditions meningitis may run a perfectly symptomless course, or occasionally its symptoms may be obscured by the symptoms of another (primary) disease it is at once understood how inestimably we are indebted to Quincke for introducing lumbar puncture as an aid to our diagnostic methods. We shall consider the subject of lumbar puncture under the separate forms of meningitis. While the acute inflammatory changes which occur in the meninges may be serous or purulent, they do not differ essentially; although certain infective agents favor the production of one or the other variety. It is important to note that for purulent meningitis to supervene, an interval of considerable time between infection and death must have elapsed.

When, in consequence of the powerful toxic effect of the infective agent, death occurs within a few hours, we find merely a serous exudate, notwithstanding that the infective factor may have extremely potent pyogenic properties.

The difficulty of recognizing the earlier stages of the inflammation in the cadaver makes it comprehensible that the purulent varieties and those beginning with copious serous exudation (as, tuberculous meningitis) have been known longer and are better understood than those in which only a slight serous moistening and delicate microscopical changes of the leptomeninges and of the cerebral cortex are found. And yet these very cases have recently been so zealously studied and have attained such clinical importance, that their discussion in this chapter is imperative. The various avenues and modes of infection will be discussed under the separate captions.

DISEASES OF THE DURA MATER

Only two diseases of the dura mater are important from a clinical standpoint, meningeal apoplexy and internal hæmorrhagic pachymeningitis.

Meningeal apoplexy embraces the hæmorrhages of the blood vessels of the dura.

As these usually are the direct consequence of severe cranial traumatism, frequently accompanied by concussion of the brain, a description of their clinical symptoms here is manifestly unnecessary. The methods of diagnosing and treating these hæmorrhages can be found in the text books on surgery. However, one point merits special mention. After difficult delivery *hæmorrhages of the dura mater* are frequent. Depending upon their extent they either terminate fatally within a few hours or days, with the symptoms of cerebral compression, or excite temporary manifestations of irritation, or run a course with no immediate symptoms to sometimes induce gradual cerebral changes; when this occurs the symptoms which ensue belong to the group of infantile cerebral paralyses. Moreover such pachymeningeal hæmorrhages are associated with similar hæmorrhages of the pia.

Internal hæmorrhagic pachymeningitis now appeals to the pediatrist inasmuch as it has become diagnosible and amenable to treatment by lumbar puncture. The anatomical sequence in this disease is as follows: during the first year of life, in children debilitated by malnutrition, syphilis, rickets, the infectious diseases, etc. (scurvy), usually as the result of a somewhat milder injury to the skull, an exudation from the inner lamella of the dura supervenes, which forthwith constitutes serous external hydrocephalus. Pachymeningeal hæmorrhages into this fluid follow from time to time, in consequence of which the cerebrospinal fluid becomes tinged with blood. This clinical picture, a detailed description of which we may omit because, apart from the absence of fever, it is practically the same as that of acute leptomeningitis, is dominated by the symptoms of brain pressure recurring periodically (bulging of the fontanelles, congestion of the papillæ, retinal hæmorrhages, vomiting, pulse anomalies, and hydrocephalic stare).

Cerebral compression diminishes as the exudate becomes organized, but within a period of days or weeks returns and, if untreated, death ensues within a few weeks or months, often after hydrocephalus has developed to such an extent as to be visible externally.

The **diagnosis**, when the symptoms mentioned above are present, is possible only if lumbar puncture discloses fluid under increased pressure and uniformly tinged with blood; provided the presence of blood, due perchance to the puncture can be excluded. In such a case the diminution of intracranial pressure wrought by the puncture may induce prompt amelioration of the condition and a quicker absorption of the exudate, without fresh hæmorrhages resulting from it. Herein lies the possibility of a permanent cure. It is unnecessary to state that this procedure must be supplemented by careful nursing and diet.

DISEASES OF THE PIA MATER

With reference partly to the pathologic picture, partly to the nature of the etiologic factor, we differentiate among the acute and subacute leptomeningitides, the following forms: first, tuberculous meningitis; second, purulent meningitis; third, so-called cerebrospinal meningitis (genickstarre). While the first and third of these varieties are generated by definite infections, purulent meningitis can be caused by all possible microörganisms which exhibit pyogenic properties. Certain clinical courses correspond to these bacteriological types, although isolated observations often do not disclose the basic type.

We shall begin with the description of tuberculous meningitis or basilar meningitis (meningitis basalis).*

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* The term "M. basalis" is widely employed, especially in English and American literature, to designate also nontuberculous (e.g. pneumococcic) meningeal inflammations located at the base of the brain. As there can be no logical criticism of this use, and further, since the involvement of the convexity often preponderates in tuberculous meningitis, it is better always to designate the tuberculous form as such and not as basal.

TUBERCULOUS MENINGITIS

Tuberculous meningitis is invariably caused by Koch's tubercle bacillus.

The insignificant pyogenic faculty of the tubercle bacillus is recognized; hence the probability of a *mixed infection* is suggested whenever, as not infrequently occurs, considerable pus is encountered in a case of tuberculous meningitis. Mixed infection may be caused by a number of microörganisms, most frequently the meningococcus and the pneumococcus. It apparently does not have much influence on the course of the disease; its effect on the pathologic process and especially the cellular constituents of the cerebrospinal fluid is greater.

While clinically, tuberculous meningitis often produces the impression of an absolutely independent disease, since it attacks children who have hitherto been healthy, nevertheless it must be maintained that it is always a secondary disease.

The *primary tuberculous focus* may be situated in the immediate vicinity of the meninges, in the nasal cavity (Demme), in the bones of the spine or skull, in the middle ear and in the mastoid cells. The tubercle bacillus forces its way along the lymph channels to the meninges, and if the conditions are favorable remains there and multiplies. Occasionally also the bacillus can reach the cerebrospinal canal by way of the lymphatic investments of the nerves, a mode of infection to which Strümpell and Leube particularly have directed attention.

As a rule infection by extension appears to be of much rarer occurrence than by metastasis through the circulation of the blood.

An argument in favor of this is the arrangement of miliary tubercles along the vessels of the pia mater, sometimes in such a manner that the terminal expansions of only a single branch or of some rather diminutive branches are attacked. The primary foci, corresponding to the prevailing character of tuberculosis in childhood, are situated chiefly in the lymph-nodes. Of these the cervical and submaxillary nodes, which receive the lymph from the mouth and pharnyx, and especially the bronchial and mediastinal nodes are probably the most frequent sources of infection, indirectly however by way of the blood vessels. The breach through which the tuberculous virus enters the circulation is by no means always discoverable in the cadaver; but it has been found so often (by Huguenin before Weigert's work had directed attention to this point, and later by others) that this method of infection is positively established.

Besides the lymph-nodes, of course pulmonary or osseous or urogenital tuberculosis can furnish the primary focus. Almost always miliary tuberculosis of the liver, spleen, etc. (often however so slightly developed as to preclude clinical diagnosis) is found at the autopsy. Just as miliary tuberculosis in other non-nervous organs in cases of meningitis is almost invariably found, so there are, on the other hand, numerous cases in which, notwithstanding widely disseminated miliary tuberculosis of the entire organism, the pia mater has remained intact. This fact presents one of the most interesting problems of the pathogenesis of meningitis.

The dissemination of tuberculosis in the body proceeds so irregularly that it is impossible to await or predict its seizure upon the meninges at a definite stage. For this reason, search for an exciting cause for this event is the more justifiable—even if we do not reach a positive result.

Etiology.—Here we may place in the front rank the infectious fevers, especially measles and whooping-cough. They, like all *exciting causes*, act by stimulating a localized, more or less dormant tuberculous process. But in regard to whooping-cough it must be observed that it is often a question whether the spasmodic coughing spells have not indeed been the expression of advanced tuberculosis of the bronchial glands. It appears more certain that measles may incite a hitherto latent glandular tuberculosis to a more rapid miliary dissemination which may involve the meninges.

The validity of trauma, at least of ordinary blunt injuries to the skull, as an exciting cause is open to argument. Of course, if a tuberculous focus is pierced or crushed, this may result in starting a general miliary tuberculosis with meningitis. We observe this after forcible repression of a spondylitis or tuberculous coxitis, after operations on the cervical glands, etc.

Less certain is the inciting part played by cranial injuries, striking the head against level ground, falling from a bed or chair, bumping the head on the furniture, etc., if no cerebral symptoms of any kind immediately follow the accident itself. At any rate to assume this connection as established, it is requisite that the interval between the injury to the skull and the onset of meningitis should correspond approximately to that which is required for the experimental production of tuberculous meningitis in animals.

Robert Koch found that eighteen to nineteen days elapsed after intravenous injection before a microscopically discernible miliary tuberculosis appeared; L. Martin found the time was shorter or longer, depending upon the nature of his research animals and the concentration of his cultures. From this experimental evidence we must conclude that in those cases in which death follows the injury within a few hours or days, the injury merely offers an opportunity for a meningitis which was already anatomically present to come into clinical view. We shall however accept the traumatic etiology in those cases in which at least eight to ten days, usually indeed three to four weeks, intervene between the cranial injury and the first appearance of meningeal symptoms. Observations of this kind have been repeatedly described and we ourselves have very frequently made them with especial attention to this point. Death from meningitis occurs so remarkably frequently about the third or fourth week after a fall or a blow that it is impossible to consider it merely a coincidence.

In comparison with infectious disease and traumatism, the other suggested supplementary causes of tuberculous meningitis, such as sunstroke, mental overexertion, violent mental excitement, etc., play at most a vague and secondary part.

It should be mentioned that chronic nontuberculous otitis media can very frequently be demonstrated in individuals afflicted with tuberculous meningitis and no doubt is a predisposing cause. The digestive disturbances which sometimes precede meningitis, especially in young, otherwise healthy children are to be esteemed rather as prodromal symptoms than as the cause of the disease.

Pathology.—The most striking meningeal changes are the miliary tubercles and the inflammatory exudate; corresponding to these in the ventricles, the granular ependymitis (which however is only partly tuberculous), and the internal hydrocephalus. The changes in and on the vessels, not discernible macroscopically, are important, since they cause more or less circumscribed circulatory disturbances in the brain, and their resulting symptoms.

The tubercles are found chiefly at the base of the cerebrum and cerebellum, especially numerous within the region covered by the artery of the Sylvian fossa, somewhat less frequently and less numerous on the convexity. The fact that the miliary tubercles are arranged along the ramifications of the medium-sized and smaller arteries has always appeared a support for the theory of leptomeningeal infection through the blood. Although this assumption seems natural, yet it should not be forgotten that the same arrangement exists also in those cases in which a direct infection of the subdural and subarachnoidal fluid is much more probable.

The fact that Louis Martin found the same distribution of tubercles in his animals, after he had injected cultures through the atlanto-occipital joint into the subarachnoidal space, without injuring the vessels, agrees with the latter view. It may be assumed that the lymphatic sheaths of the vessels then undertake the distribution, probably because they are wider or possess stronger currents than the other lymph-spaces.

The inflammatory exudate, of a peculiar gelatinous consistence, is located chiefly at the base of the brain between the optic chiasm and the cerebral peduncles. By virtue of its serofibrinous consistence it adheres quite firmly to the meshes of the pia mater and, when the brain is removed, does not run out like serous inflammatory œdema which often is much more widely disseminated.

By more or less abundant admixture of cells the exudate, which in a pure state is clear, is often tinged gray or yellowish green, at times even resembling pus. The bands of exudate enveloping the vessels often extend from the base to the convexity, and the sheaths of the cranial nerves also appear already macroscopically infiltrated.

We shall return later to the cellular elements of the exudate and cerebrospinal fluid, which are important in relation to clinical diagnosis.

As evidence of the participation of the cerebral ventricles in the inflammatory process we find granular ependymitis (apparently possessing no physiologic significance), the chorioid plexus infected with miliary tubercles and exudate, and—most important—internal hydrocephalus.

Its development may show decidedly different degrees if we take as a criterion, on the one hand flattening of the cerebral convolutions, on the other, size of the ventricular cavities. While compression phenomena, like the symptoms of cerebral pressure, never fail to appear in tuberculous meningitis, the hydrocephalus may in one case be slight, in another case so considerable as to justify the suspicion that an abnormally large ventricular system already existed previous to the meningitis.

The significance of this symptomless stationary internal hydrocephalus, since it is so frequently combined with tuberculous meningitis, would probably be that it represents the cause of or indicates a pathological condition of the central nervous system which facilitates the infection.*

A critical study of the circulatory and nutritive relations of the central organs might afford a decidedly interesting explanation of this complicated matter.

Clinical Discussion of Tuberculous Meningitis.—Tuberculous meningitis, as we have already stated, presents a secondary localization of the tuberculous infection. The few observations in which the primary focus could not be discovered do not invalidate this fact. As will be shown later, however, the clinical manifestations of the disease are often influenced by the association with advanced tuberculosis of other organs. It is important to consider the relationship of tuberculous meningitis to other tuberculous diseases. The available statistics (Brandenburg, Müller, Haushalter and Fruhinsholz, and others) based upon post-mortem material, supply no perfectly satisfactory picture, and we accordingly refrain from discussing them. Our own clinical experience allows us to state the following:—

In the course of widespread pulmonary tuberculosis in children we observed tuberculous meningitis supervene just as infrequently as is the rule in adults, even if, as often happens, a recent miliary tuberculosis which had caused no independent clinical symptoms was found at the autopsy.

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* Our own researches, which as yet have not succeeded in clearing up these relations, have taught us that when the ventricles are enormously dilated, quite regularly there is found a hypoplasia of the suprarenal substance such as Czerny has described in pronounced "idiopathic" hydrocephalus.

Tuberculosis of the peritoneum, abdomen, genitalia or bones, leads to meningitis almost as infrequently, except indirectly through miliary processes widely distributed and clinically plainly recognizable. The experience, repeatedly corroborated in the literature, that meningitis very often succeeds operative interference with tuberculous lymph-nodes, joints, or bones, agrees with this observation. Here the exciting of a quiescent focus leads to numerous metastases, which involve also the meninges, exactly as occurs in other cases through the agency of acute fevers, traumatism, etc.

The younger the meningitis patients are, the more frequently are cases met with in which there was suspicion of latent tuberculosis before the appearance of the prodromal symptoms. These children may be attacked by the disease while in perfect health, indeed many of them appear exceptionally robust. Henoch's dictum that tuberculous meningitis is a terminal localization of tuberculosis should not lead to the error of awaiting meningitis only in severely tuberculous individuals.

The diagnosis of tuberculous meningitis, like that of all internal tuberculous infections, is facilitated by the knowledge of a *hereditary taint.* Tuberculosis of brothers and sisters or the statement that a parent belongs to a tuberculous family and has shown positive or probable symptoms of this disease is often of decided significance. We can literally subscribe to the statement of Lederer:

"Tuberculous meningitis is often a subtle reagent upon a tuberculous taint in a parent, like hereditary syphilis upon a former syphilis in an ancestor. In both cases a descendant betrays the hitherto concealed history. There are certain families in which one or both parents when young positively had even advanced tuberculosis, and in whom the process was cured or arrested, so that afterwards they became so robust and vigorous as to surprise former acquaintances. Not these qualities are transmitted to their offspring, but the predisposition to tuberculosis with its manifestations in childhood."

It is our impression (which it is difficult to prove statistically) that those parents with reported former tuberculosis or with a latent tuberculosis more frequently lose one of their children through tuberculous meningitis; while tuberculosis of a child of parents afflicted with florid phthisis appears in the majority of cases as pulmonary tuberculosis also, and terminates fatally without meningeal sequelæ. Perhaps the magnitude of the infection is the decisive feature in these cases.

Since *mental overexertion and excitement* are frequently mentioned as factors in the etiology of tuberculous meningitis, perhaps it is not superfluous to emphasize that restless and excitable or precocious and emotional children have not, in our experience, exhibited any increased predisposition.

Sex has no influence—which could not be accounted for by statistical errors—though it is maintained that boys are more frequently attacked than girls.

Age is undoubtedly an etiologic factor. According to Huguenin the greatest predisposition exists from the second to the beginning of the seventh year; there are fewer cases from the sixth to the tenth year and the frequency diminishes rapidly from the tenth to the fifteenth year.

More elaborate statistics than these do not help us, since they depend too much on extraneous circumstances; however the question how early tuberculous meningitis may attack a child is of interest in view of the still obscure requirements for and modes of infection.

Congenital tuberculous meningitis and meningitis during the first week of life have never been reported. It has been observed by Steffen, Henoch and ourselves in children who died at the age of three months. Hohlfeld and Finkelstein repeatedly saw cases in early infancy. Dr. Hubert Reich (the district physician of Mühlheim) in 1878 reported a very remarkable series of cases in early infancy. He saw ten infants of healthy families die of tuberculous meningitis within fourteen months. The cases occurred in the practice of a midwife who had severe phthisis, and who was accustomed to inspire air from mouth to mouth in cases of asphyxia and to frequently kiss upon the mouth all the children at whose birth she assisted. One child died at the age of two and a half months, seven between three and four months, one at nine and one at six months. As all of these infections (except perhaps the two last) probably occurred directly after birth, from bacilli in the oral secretion of the midwife, the observation that meningitis terminates fatally at the earliest two and a half, in the majority of cases three to four months after infection, is certainly of great importance. The disease lasted from one to three weeks in all the cases, but, so far as could be ascertained, was preceded by bronchitis, emaciation, anorexia, etc.

Symptoms.—The symptomatology and course of tuberculous meningitis are so varied that it is impossible to advance a valid type of all or even of the majority of cases. Mention has already been made of the easily comprehensible influence which disseminated tuberculosis, the general condition of nutrition, secondary diseases, and, finally, exciting trauma, may exert.

Hence if we endeavor to arrange a scheme of the disease, we must for the present omit all cases appearing at the end of severe general tuberculosis or cachexia of other origin and all fulminating cases subsequent to severe injury.

Robert Whytt who first described tuberculous meningitis, distinguished definite stages in the course of the disease.

As subsequently modified and accepted by most authors Huguenin has formulated them as follows:

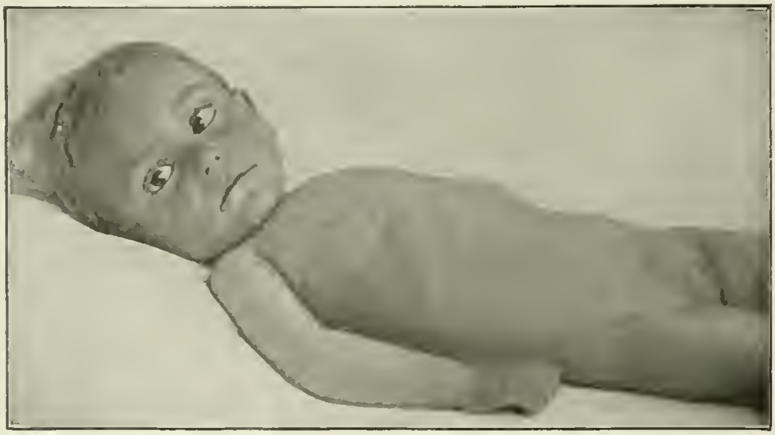
I. *Stage of Cerebral Irritation.*—Headache, vomiting, constipation, retraction of the abdomen, incipient cephalic pulse, excitation and delirium, convulsions in children.

II. *Stage of Pressure.*—Pupillary symptoms, soporose and comatose conditions, cephalic pulse, constipation, retracted abdomen, cervical rigidity, facial paralyses (generally referred to the base of the brain), lesions of the facialis, hypoglossus, oculomotor and abducens nerves, hemiphlegia, contractures, convulsions; frequent change in general health.

III. *Stage of Paralysis.*—Coma, immobility, diminution of all spasmodic symptoms in general, increase of paralyses, disappearance of cervical rigidity and retraction of the abdomen, irregularity of pulse and rapid increase in pulse rate, agonal rise or fall of temperature, death.

Owing to the impossibility of sharply dividing these stages, we shall follow the precedent of Huguenin and others and reject them as a basis.

FIG. 64.



Child ten months old. Somnolence, left internal strabismus (abducens paralysis resulting from basal exudate). The plate exhibits the fixed, vacant expression of meningitis.

Meningitis (in children hitherto healthy or not yet very sick) begins, in the majority of cases, with certain *prodromal symptoms,* which in themselves are not pathognomonic of meningitis, but if several occur together, should arouse suspicion.

For example, a child two to three years of age begins to lose interest in play, becomes moody, and all the efforts of its family to please it scarcely succeed. At times it is peevish, it loses its appetite and frequently, but by no means always, vomits.

There is usually, at an early date, disturbed, broken sleep, lighter than in health.

As a rule in this stage physical examination discloses nothing definite; at most the tongue is somewhat coated and the temperature is slightly elevated—to 38° C. (100° F.) or a little higher; pulse and respiration are normal or, in the presence of fever, only slightly hurried. Therefore the attack is often regarded as a digestive disturbance, an error which even the experienced observer cannot always avoid. But one should not, as often happens, because of the normal stools, speak of a gastritis, as this presents no independent symptom-complex. Doubt concerning the diagnosis does not as a rule persist longer than two or three days; then the failure of the prescribed treatment (laxatives, scant diet of cereal waters) indicates the serious nature of the disease. It is evident that the child grows constantly weaker, more apathetic, scared. He sits by his toys for a long time without playing. At times he lies down and would gladly go to bed also in the daytime. He lies there half awake or perhaps falls asleep and arises after one or several hours apparently refreshed. But the improvement does not last long; play is not very alluring. At times the child sighs, but to the question whether anything hurts, even children four to six years of age seldom say definitely that they have headache; more frequently they complain of being tired, pains in the legs (in the tibiæ) or abdomen.

Physical examination even at this time usually discloses an indefinite pulse anomaly, which with the advance of the disease soon becomes more evident and is numbered among the most constant symptoms of meningitis. The pulse is irregular and uneven, especially after moving about, changing position and the like. The pulse rate is not altered in the same manner in all cases; it is usually normal, seldom somewhat increased (corresponding to the low fever) occasionally a little diminished. Increased tension and pulsation of the membrane of the fontanelle, in children with patulous fontanelles, indicate the commencement of increased intracranial pressure. In older children exaggerated tendon reflexes and hypertony of the muscles of the extremities appear as its symptom.

Very soon the child takes to bed, never to rise again. The clouding of the mind becomes protracted and deeper; sleep is not sound and quiet, but the child often lies half asleep, with half-closed eyes, nearly always with ruddier cheeks than it had in the months just before the attack. Its head and brow, occasionally the whole body, are often moist, as is the case in a sleeping healthy child. This dozing is interrupted at variable intervals by monotonously repeated movements, as picking the lips or bed-spread, by peculiar sucking or chewing motions, by deep, sighing inspirations or short piercing shrieks ("*cris hydrocephaliques*"). Great fright at sudden noises, sudden brilliant illumination, or touch indicates that besides somnolence, there exists hyperæsthesia of the sensory organs. From a slight stiffness of the neck at first, even at this time there succeeds firm retraction of the head, so that it bores into the pillow.

Increasing hypertony of the muscles with heightening of the tendon reflexes accompanies this feature.

The abdominal muscles are contracted causing the "boat shape" appearance. But at the outset it should be stated that this symptom, so significant in older children, is often lacking in infants; on the contrary it is often replaced by a decided distention of the abdomen. The same holds true of constipation, which in older children is almost constant and of higher degree than can be explained by the reduced amount of food taken; in its place we much more frequently find in infants, at least in those artificially nourished, severe diarrhœa. In this advanced stage of the disease vomiting is much rarer than in the beginning.

The behavior of the *temperature* does not follow any fixed rule; it may be subnormal, normal or elevated; indeed it may be elevated in any stage or form of the disease.

The *pulse*, usually retarded to 70–60—in infancy to 100–90—beats a minute, is likewise uneven and irregular. The instability of pulse rate, mentioned as an early symptom, its jerky racing, caused by the patient sitting up or by painful passive motions, usually persists, to be displaced after some days by a constant bradycardia.

Respiration is often irregular, sighing, but not pronouncedly periodic.

The *pupils* are usually contracted, often uneven and either do not react or react slowly and incompletely to light. Usually the half open eyelids disclose the eyeballs now rolling irregularly, now in conjugate deviation, now making vertical nystagmatic motions. At the same time there is usually a severe pericorneal conjunctivitis in the form of vessels extending to the limbus.

Constantly deepening *obscurity of consciousness* is the next evidence of the progress of the disease. The patient lies in deep coma, unable to swallow. The head has ceased boring into the pillow; the extremities hitherto rigid or shaken by clonic spasms, gradually relax; in consequence of inability to cough, the respiration becomes audible, rattling, and as the end approaches often assumes a periodic character; the pulse always becomes rapid (though the earlier bradycardia in not a few cases may have been absent); the temperature rises during the last few days or hours to 40° C. (104° F.) and higher, or it may become subnormal. The sweat-glands, formerly active, cease to functionate; the skin is pale, often grayish, or brownish, dry, desquamating, and owing to the rapid emaciation it can be lifted in folds. In deepest coma, with penetrating tracheal râles and thready running pulse death finally ensues.

The **duration** of the disease, in ordinarily typical cases, is about three weeks, but these limits vary widely, as we shall immediately note in portraying the chief variations.

These *deviations* involve the mode of onset as well as the combinations of symptoms and additional symptoms, and the entire course of the disease, which occasionally is protracted by considerable remissions or other circumstances. The onset, usually insidious, as we have described it, may be sudden, without prodromal symptoms. Not infrequently it is ushered in by convulsions, which either resemble Jacksonian epilepsy (in which case after continuing for minutes or hours they are followed by paresis or paralysis of the extremity first attacked), or they resemble genuine epileptic attacks. Tuberculous meningitis begins with tonic spasms much less frequently than the other forms of meningitis and, especially in infancy, so rarely in comparison with the frequency of clonic convulsions that the occurrence of general clonic convulsions in infancy justifies the suspicion of tuberculous meningitis only in the presence of more definite symptoms.

The early occurrence of *convulsions* in tuberculous meningitis depends often, if not always, upon an unusual involvement of the convexity. As a rule this involvement is asymmetrical, being limited to one hemisphere, and in this to the area supplied by the arteria cerebri media. In such cases the meningeal exudate may in circumscribed localities, preferably the motor area, attain a striking thickness and firmness (through its richness is fibrin) which explains the focal symptoms (convulsions and paralyses). This form of anatomical deviation is known as "méningite en plaques." Further, these focal symptoms are often very transitory, in which respect they differ materially from the more constant symptoms of tumors. Convulsions or paralyses in different localities not infrequently interchange with or succeed each other; and then by recalling the topographical arrangement of the cortical motor areas, we can recognize on the one hand the localization of the irritating process on the cortex, on the other hand its transitory nature (circulatory disturbances, œdema, etc.) (Zappert).

From this last point it is comprehensible, and this should be especially emphasized, that the same clinical symptoms occasionally may occur also without a previous "méningite en plaques," and that therefore our diagnosis does not extend beyond a certain probability.

The particular cases described exhibit in their further course, the greatest conceivable difference. The course may be very stormy and contrary to the rule rush through the separate stages to death within a few days; however more frequently it is abnormally protracted. The headache following the initial convulsions and the changes of consciousness may disappear; the paralyses, if any existed, diminish, and then after days or weeks the regular picture of tuberculous basilar meningitis first begins to develop. Or definite meningeal symptoms, headache, clouding of the senses, cervical rigidity, pulse and pupillary phenomena, fever, etc., follow the initial insult but the condition suddenly and surprisingly improves and a remission of a week or month or exceptionally, even a year occurs, which simulates a complete and permanent recovery.

Undoubtedly the length of time the disease lasts is especially influenced by two circumstances, nourishment and increase in intracranial pressure. This explains why the course of the disease is often extraordinarily slow in breast-fed infants, since, in the first place, suckling at the breast, which is the result of a deep-seated reflex mechanism, becomes interfered with very late and, in the second place, the yielding cranium keeps the increasing cerebral pressure below the fatal limits for a long time. So the disease is occasionally seen to drag on for four to six weeks or longer, with a comatose stage persisting for weeks.

On the other hand, individuals in whom meningitis presents the termination of an extensive general tuberculosis, usually succumb a few days or a week after the onset of the disease. The gradual development of the disease, as we have already remarked, is usually absent in these cases; coma sets in early and dominates the symptoms of motor irritation. The development of tubercles on the meninges is usually trifling in comparison with the amount of gelatinous exudate.

In conclusion those atypical cases are to be mentioned in which *spinal symptoms* are more prominent than usual. While we are well aware that participation of the spinal meninges is the rule (although perhaps less frequent with miliary tuberculosis than with nonspecific inflammatory processes), still there may be no clinical signs of it. Exceptionally however we observe severe radiating pains in various segmental divisions and early loss of the tendon reflexes, which indicate disease of the nerve roots; and occasionally undoubted retention of urine, ischuria paradoxa, which is not caused by the dulness of the senses and demands artificial evacuation by expression or catheterization.

Having thus briefly sketched the atypical forms (it is impossible to exhaust the variety of possibilities), we shall discuss the individual symptoms to complete what has been said.

The *temperature* follows no definite curve; measuring it twice daily it may be found normal for a long time or even throughout the attack or, especially in young infants, it may become subnormal shortly before death. In other cases there may be continuous fever similar to that of typhoid; most frequently, however, there is a low fever, 38°–38.5° C. (100°–101° F.) often slightly and irregularly remitting, which in the last days of life first shows gradual a rise to 40°–41° C. (104°–106° F.) or higher and which probably is due to complicating bronchitis and bronchopneumonia.

Frequently a fall in temperature occurs with the onset of meningitis if a disease with high fever, *e.g.*, advanced pulmonary tuberculosis, preceded the meningitis. The cause of this behavior is not apparent but it should be called to mind that frequently purulent processes in various parts of the body, *e.g.*, ulcerating lymph-glands, etc., also severe weeping eczemas, tend to subside quickly under the influence of meningitis. Probably a similar process occurs when pulmonary tuberculosis and the like are concerned in which a mixed infection no doubt causes the high fever.

The behavior of the *pulse* is of far greater diagnostic value than the temperature curve. The previously mentioned irregularity of the pulse as a rule begins early and is seldom absent; it is, however, in itself, not pathognomonic of meningitis, since an identical pulse occurs occasionally with digestive disturbances and other non-cerebral diseases. But when there is associated with this irregularity a palpable slowing of the pulse, in connection with other symptoms, the strongest suspicion is justified. The diminution of pulse rate may be very pronounced, to 60–50 beats a minute and may persist continuously for many days: as a rule however as indicative of the great instability of the pulse, there occurs a transitory jerky rise in the rate on change of position or with painful irritations. The stage of bradycardia does not invariably occur. This is especially the case in children less than a year old and is probably accounted for by a still faulty action of the inhibitory vagus fibres at this age. As the case advances the slow pulse uniformly disappears being replaced by tachycardia (160 to 200 pulsations per minute), which indicates vagus paralysis, and persists till death. The pulse rate seems to be absolutely independent of the temperature.

Respiration is much less characteristically influenced by meningitis than is the pulse. It may early become irregular; after a deep noiseless inspiration and its succeeding sighing expiration, a lengthened pause ensues. But we see this also in other patients, whose minds being disturbed, suffer pain. The periodicity of respiration which occurs as the end approaches is more significant. The periodicity may be of the pure Cheyne-Stokes type; it may however be such that a series of equally deep respirations is suddenly interrupted by a considerable pause, in which case the gradual ascent and descent characteristic of Cheyne-Stokes respiration is absent. This type is called meningeal respiration.

A periodic variation of the size of the *pupils* is associated with the periodic respiration. The pupils are of ordinary size or at most only slightly contracted during the respiratory pause, while, synchronously with the beginning of respiration they slowly and widely dilate. With the cessation of breathing both pupils return to their former size quicker than they dilated, to repeat this play with the next respiratory phase.

Intense light during the respiratory pause produces an observable, although not the maximum reflex contraction; but, during the progress of the respirations, it does not prevent the increasing dilatation of the pupils. Pinching any part of the body does not induce such a dilatation of the pupils as would disturb the periodic changes in the size of the pupils (Thiemich).

The pulse rate also is influenced by the periodic respiration; from the beginning of the first inspiration to the end of the pause it constantly decreases.

Sensory disturbances form an essential part of the symptom-complex. An early sign of this, even before pronounced lethargy ensues, is diminished frequency of winking, which depends upon lessened sensibility of the cornea.

From the beginning the *psychic change* as a rule is of a depressant nature; at any rate a stage of excitation precedes it much seldomer than *e.g.* in cerebrospinal meningitis. Usually there is deep coma for days, but occasionally it may set in only a few hours before death. Again in other cases it may occur early and may persist for weeks until the end.

Headache continues even in the presence of marked somnolence and betrays itself occasionally by tossing, painful sighing, or by abrupt, shrill cries; this so-called "cri hydrocéphalique" is however neither a constant nor a positive symptom, as it occurs also in other conditions of cerebral irritation.

Constipation, which is a quite constant symptom, though somewhat oftener absent in artificially nourished infants, may be due in part to insufficient ingestion of food (which is evidenced by extreme emaciation and shrivelling of the patient); in part it has a centric origin as has the boat-shaped abdomen.

Vomiting, which was mentioned as a frequent early symptom, is certainly centric. It only remains to emphasize that the vomiting by no means always has the projectile character, which is held to be characteristic of cerebral emesis, and that conversely, this type itself frequently occurs in gastric diseases, *e.g.* in spastic contraction of the pylorus.

Of changes in the *skin* perspiring and erythematous patches especially if these appear after lightly touching or gently stroking the skin, deserve attention as being signs of vasomotor disturbances. The patches described above were named by Trousseau "tâches cérébrales."

Disturbances of function of the *cranial nerves,* paralysis of the facial, hypoglossal and oculomotor nerves in varied combinations and sequence, usually occur comparatively late as a result of the basal exudate. This holds also for the pupillary disturbances which, at least in the later stages, belong to the most constant symptoms: inequality of the pupils (anisocoria), slow and incomplete reaction to light, later complete reflex rigidity of the pupil; the pupils may at this time be strikingly dilated as well as extremely contracted (Schlafpupillen). Pinching any part of the body then no longer excites dilatation (Parrot).

Recently Squires has asserted that, on the fourth or fifth day of the disease, there occurs a constant mydriasis on bending the head backward, a constant myosis on bending it forward. There is as yet no corroborative evidence on this probably important symptom.

Besides *optic neuritis,* which is rather infrequent and not very valuable for diagnostic purposes, the occurrence of tubercles in the choroid should be particularly mentioned. Its positive recognition, which is not a simple matter (Schmidt-Rimpler), may be decisive in suspected cases; owing to its infrequency its diagnostic value is decidedly limited.

Lumbar puncture, which discloses to us, on one hand the existence of pressure in the cerebrospinal canal, and the state of the cerebrospinal fluid on the other, is the most reliable method we possess for positively establishing the diagnosis of tuberculous meningitis.

The pressure symptoms, which Pfaundler especially has studied, show a certain definite dependence upon the age of the patient, the chief localization of the meningeal process, and especially the stage of the disease. The pressure, normally averaging 20-25 mm. Hg., rises during the disease to 18–52 mm., and declines again to normal with the appearance of cerebral paralyses. Of course, decided variations occur in individual cases; still at the height of the disease increased intracranial pressure can always be detected. The increase of albuminoids, diminution of reducing substances, etc., are of subordinate diagnostic value. The following points are more important:—

1. The cerebrospinal fluid, which is altered by inflammation, on standing, precipitates a *fine fibrinous coagulum* resembling a spider web; which on agitation is loosened from the walls of the tube and rolls itself up into small floccules. As the same thing occurs in some nontuberculous serous meningitides, it is differentially diagnostic only as against functional disease, but not against other forms of meningitis.

2. The fluid is absolutely clear much less frequently than is usually reported; in most cases it shows rotating turbidity, like dust in sunlight: and as the disease progresses it becomes plainly opalescent. The precipitate always contains some cells, in which *mononuclear lymphocytes preponderate*. This difference from the other meningitides, which had already been established by Bernheim and Moser, was subsequently studied by Widal and his collaborators and promulgated in numerous publications as cytodiagnosis. We have reason to emphasize the priority of the German authors.

The determination of the pure or preponderating lymphocytosis of the fluid is valuable for the diagnosis of tuberculous meningitis: it is corroborative but not pathognomonic. A positive diagnosis can be made only by the discovery of *tubercle bacilli*. While this is not easily accomplished, the search is facilitated by examining the fibrin coagulum which contains the bulk of the cells and bacteria; the longer and more carefully the search is continued, the oftener is it successful. The chief difficulty consists in the scarcity of bacilli especially at the beginning of the disease. Therefore Langer has proposed incubating the aseptic puncture fluid, in which within two weeks at most a proliferation of bacilli with the formation of a deposit resembling bread crumbs occurs. Of course this slow method is useful only in cases in which there can be no postmortem.

Our present knowledge of acid-resisting bacilli renders it desirable to employ, for identification, culture and animal experiments besides microscopical examination; at least in those cases which, because of recovery, justify extreme skepticism.

The **prognosis** of tuberculous meningitis is almost absolutely fatal. Exceedingly few cases have been cured, at any rate cases in which tubercle bacilli were found in the cerebrospinal fluid; and none of these were kept under observation for years, as is requisite, in view of what has been stated regarding long-continued remissions. Barth's case is conspicuous through the fact that after the child had been mentally blind and deaf nearly a year, it made a complete recovery.

Treatment.—As yet there can be no discussion of a therapy for tuberculous meningitis. Of course the physician should treat the symptoms of irritation particularly and not spare narcotics in case of necessity; but this question arises much less frequently than in meningococcus meningitis. Headache is often temporarily relieved by lumbar puncture; more is not attained by this procedure. In the few cured cases no methods were employed which had not proven futile in numerous others. We note among these, besides repeated lumbar punctures, copious depletion by leeches on the mastoid process (for eight days), and large doses of creosote. The major operations on the skull which, repeatedly tried on account of the analogy to tuberculous peritonitis, promised success have so far always resulted unfortunately; and it is still doubtful if the expectation (Sokolow) still cherished regarding this method will ever be realized.

PURULENT MENINGITIS

(Meningitis simplex)

Purulent meningitis differs both anatomically and clinically from tuberculous meningitis, at least from typical cases with wide dissemination. In the former the onset is sudden, with high fever and extensive participation of the cerebral cortex; in the latter the onset is insidious, with slight fever, a duration of weeks and a preponderance of basal exudation. In the former type there is purulent, in the latter gelatinous, infiltration of the pia mater.

Etiology.—These characteristics justify the classification of purulent meningitis as a separate group, but it is to be noted that it represents no etiologic entity. All known pyogenic organisms may cause it. Arranged with reference to their frequency these are: first, the pneumo- and streptococcus; second, the staphylococcus pyogenes (usually aureus) and Friedländer's diplobacillus; lastly, the bacilli of the coli group, the bacillus pyocyaneus and Pfeiffer's bacillus hæmophilus (influenza bacillus). The discovery of a definite microbe as yet has only scientific interest; but it appears as though the course and also the prognosis (within limits) are dependent upon the nature of the infective agent.

Purulent meningitis is without doubt more frequent in early childhood than later and, unlike tuberculous meningitis, does not spare the newborn. The protected position of the central nervous system, the investments of which nowhere reach the surface of the body, explains the rarity of primary or at least of cryptogenetic purulent meningitis. Apart from cases of general pyæmia it originates by extension of an inflammation in the vicinity of the brain or spinal cord, or by metastasis from an abscess. Examples of sources of infection belonging to the first class are the orbit, the nose and the ear. Pus may reach the pia mater by extension along the sheath of the optic nerve (seldom however) after operation on a phlegmon of the orbit with subsequent infection of the wound.

Schmidt-Rimpler reports such a case in a boy seven years of age, in whom purulent basilar meningitis ensued months after the primary injury to the orbit had healed. It is remarkable, as this author alleges, that after panophthalmia and all inflammations confined to the bulb, optic neuritis and sympathetic ophthalmia occur; but meningitis does not unless the lymph-spaces of the optic nerve have been opened by the enucleation.

It is our experience that more frequently the ethmoid bone is the portal of entrance for the germs which had previously infected the nose. Meningococci and pneumococci, which often are found in the nasal cavity in normal individuals, are probably the only cocci which can infect the meninges without clinically obvious disease of the nasal mucous membrane; however we have observed purulent meningitis in a series of infants with severe hereditary syphilitic coryza. In these cases the oldest and richest purulent foci were found at the base of the brain. This fact accords best with the assumption of an infection proceeding from the nose, although purulent otitis media, which existed in all of the cases, cannot be easily eliminated as the cause.

There is no reason to consider at this time the passages from the tympanum to the meninges which can serve as avenues for the purulent factors. But we desire to state that there is hardly any danger of meningitis in the cases in which the disease is confined to the tympanum without implicating the labyrinth. Involvement of the labyrinth is the rule in infancy. We cannot consider fully otogenetic meningitis, the peculiarities of which rarely appear in children.

We contend that *otitis media concomitans* is a thoroughly benign disease. This is the experience of clinicians and differs from the view of many pathologists who regard purulent otitis media as one of the commonest causes of death in infancy, without taking into consideration the fact that it is usually a terminal disease in a child succumbing to digestive disturbance. Without pursuing this subject further, we only add here as relevant that when, according to Ponfick ninety-one out of one hundred autopsies on children one to four years old (from the Breslau Children's Clinic) presented purulent otitis media with or without other visible organic disease, while only eight had meningitis (with or without pneumonia), the etiologic dependence of meningitis upon the otitis is not at all self-evident. It may be considered to be a fact only if an anatomical connection is shown and the otitis preceded the meningitis. Usually it can be proven that the meningitis was already fully developed before the first symptoms of the terminal (concomitant) otitis appeared. At most a secondary infection of the ear from the meninges may be assumed, by no means the converse. The otitides succeeding measles and scarlet fever appear to induce leptomeningitis, particularly by means of an infectious sinus thrombosis, more frequently than the otitis concomitans of infants to which our remarks above refer.

Furunculosis or erysipelas of the scalp leads to purulent meningitis by direct extension along the lymphatics rarely and only in cachectic children.

Infections of a septic nature (typhoid, influenza, etc.) or those localized in a distant part of the body, and which implicate the meninges by metastasis, differ from those mentioned, directly extending infections. The source of infection can very often be only surmised,—tonsillitis, osteomyelitis, etc. Purulent meningitis originating metastatically presents no anatomical peculiarities differing from cases caused by direct extension.

Comparatively frequently meningitis develops with or as the result of pneumonia or pleurisy.

It is quite certain that often in such cases the circulation carries the pathogenic germs, still the remarkably extensive participation of the spinal meninges, which we have often observed in purulent pleuritis, argues that the microbes travel by way of the lymphatics through the intervertebral foramina. The fact that at the autopsy purulent otitis media is almost invariably found associated with empyema and purulent meningitis, had led to the assumption that it furnishes the primary focus for both the other affections, a doctrine from which we must dissent for the reasons stated above.

Pathology.—The pathology of diffuse purulent meningitis calls for little discussion. Usually, besides the external meninges, the chorioid plexus and the ependyma ventriculi are diseased; indeed the purulent inflammation of the ependyma may preponderate to such an extent that the entire process might fittingly be denominated "ependymitis." It is evident that the cortex suffers circulatory and nutritive disturbances and round cell infiltration, exactly as in tuberculous meningitis, and constitutes the source of at least some of the symptoms, while some are induced by cerebral pressure.

If we now proceed to portray the clinical picture of diffuse purulent meningitis it should at the outset be emphasized that its aspect is decidedly modified by the consideration whether the disease apparently attacks an hitherto healthy child, primarily, or a child severely ill, terminally. In the latter instance, as for example with severe pneumonia or empyema the meningitis sometimes completely escapes notice, because the prostration due to the basic disease obscures the cerebral symptoms. At most the appearance of complete somnolence may call attention to the complication, but it is noteworthy that consciousness may be preserved until a few hours before death. However under these circumstances the tense, pulseless fontanelle in infants indicates cerebral involvement; in older children the startling discovery is first made at the autopsy, an experience which we have had repeatedly in cases we had watched clinically.

According to our experience the onset of purulent meningitis does not have the same influence in lowering previously existing high fever, to which we called attention as a not infrequent occurrence in the tuberculous form.

Symptoms.—The disease, in children previously not very sick, usually *begins suddenly*. With decided elevation of temperature and acceleration of pulse the child becomes restless; although there is increased inclination to sleep—during the day—the normal, sound sleep of childhood is wanting; anorexia, coated tongue and vomiting, independent of the ingestion of food are among the early symptoms. Older children complain of distressing headache and violent delirium occurs; younger children by whimpering and crying when the head is touched evince the same discomfort. *Cervical rigidity* is rarer than in tuberculous and meningococcus meningitis; but epileptiform and clonic-tonic convulsions in single limbs or groups of muscles, occur in the beginning as well as in the later stages of these much more frequently than in those forms. The abductors of the hand and the flexors of the fingers seem to us to be affected by preference. There occur small, short, slowly repeated contractions, often with associated trismus and grinding of the teeth. The convulsions recur, especially in infants, at intervals of a few minutes; sometimes tonic convulsions can be excited by striking a particular part of the body (hydrencephalic reflex convulsions). However convulsions are not an absolutely essential symptom in these forms of meningitis The sensorium appears clouded from the inception, often only slightly, usually however rapidly advancing to deep lethargy. The symptomatology and later course, apart from their greater rapidity of succession, resemble so closely those of tuberculous meningitis that we refer to its presentation and here we will select only some more important matters.

The *temperature* is always considerably elevated; doubt of the diagnosis is justified if it remains normal.

The *pulse* is less characteristic than in the tuberculous form; it is usually accelerated in proportion to the fever, especially in infants; in older children it is not infrequently retarded and irregular and shortly before death is abnormally rapid.

Rigidity and varying inequality of the *pupils* are common at the height of the disease, though by no means so constant as in the tuberculous form. The fundus is usually normal; venous stasis and neuritis are infrequent.

Disturbances of ocular motion are much commoner, resulting either from an affection of the oculomotor nerves (of which the abducens seems affected by preference) and causing a partial or complete paralysis, or as a result of cortical irritation (spastic strabismus, nystagmus, temporary conjugate deviation) or more rarely paralysis.

The functions of the other cranial nerves may be disturbed peripherally or cortically in the same manner.

From what has preceded it is obvious that certainly the course, but not the symptoms, enables us, in certain cases, to distinguish purulent from tuberculous meningitis. All other means failing, the information obtained by *lumbar puncture* is decisive: from it we learn at once whether we have to deal with actual meningitis or a symptom-complex simulating it, of which we shall speak later.

Macroscopically the cerebrospinal fluid is, in disseminated purulent meningitis, always opaque, grayish yellow or green, and on standing precipitates pus. It is moreover under increased pressure; wherefore it flows in a stronger stream, unless the cannula is obstructed by masses of pus. Microscopical examination of the opaque liquid (even without obtaining a sediment) discloses large numbers of pus cells, the polynuclear form predominating; while, as before noted, preponderance of lymphocytes is the rule in tuberculous meningitis.

To complete the examination it is necessary, at least for observations to possess scientific value, to make *bacteriologic examinations* of the sediment in stained specimens and by culture. This procedure is also proper for practical purposes in order not to overlook sporadic cases of cerebrospinal meningitis, and because the character of the pus organism has some value in prognosis (irrespective of cerebrospinal meningitis).

The *pneumococcus* is the most frequent cause of purulent meningitis; it shares, with the less frequently found influenza bacillus, the distinction of causing cases of meningitis appearing in epidemic form in limited areas; indeed it was long counted among the causes of cerebrospinal meningitis. From an anatomic standpoint it is remarkable that the meningitis caused by pneumococci develops especially in the posterior division of the base of the skull, so that many of these cases are described as posterior basilar meningitis (Fränkel, Thurston and others). This individualizes the clinical picture by the prominence of its basal symptoms, frequently protracted course and comparatively high percentage of recoveries.

Staphylo- and *streptococcus* meningitis always terminate fatally, if a diffuse process is actually present. It may be briefly noted that this form cannot always be positively differentiated during life from the partial form (especially the otogenetic). Some of the few reported cases of recovery from purulent staphylococcus meningitis obviously belong to the type last named; incorrectly diagnosed cases of meningococcus meningitis (a mistake not always easily avoided) no doubt constitute another portion.

Occasionally, in streptococcic infections, the purulent process is subsidiary to the development of gelatinous serous exudation containing innumerable cocci.

It is worthy of note that the agents enumerated above are not invariably found in pure culture, but often together or with pneumococci.

Quite frequently purulent meningitis is caused by *coli bacilli* and the *bacillus lactis aërogenes* (Scherer, Goldreich and others), which may also constitute the mixed infection in meningococcus meningitis (Sacquépée). Furthermore they do not always form pus (Concetti). This form is fatal; a single case of recovery, after lasting four weeks and without sequelæ, has been reported by Nobécourt and du Pasquier.

The *influenza bacillus* has, during the past few years, been repeatedly identified as the cause of purulent meningitis. Though the frequent marked participation of the central nervous system in the clinical picture of influenza had long before been noticed, Slavik (1899, Heubner's Clinic, in the case of a nine-months-old boy with influenza) was the first to identify, intra vitam, microscopically and culturally on blood-culture media, Pfeiffer's bacillus in the cerebrospinal fluid. While Slavik's case ended fatally, Lange (1901, Ganghofner's Pediatric Clinic, Prague) reported a recovery in a case of suppurative influenza cerebrospinal meningitis in a boy nine years of age. The short course of the disease, which began rather insidiously, is very remarkable. About ten days after the symptoms had appeared there followed, immediately after eight c.c. of purulent fluid was evacuated by lumbar puncture, reduction of fever and cure of all meningeal symptoms. It is questionable whether, as Lange surmises, the removal of this small amount of fluid, assisted the cure. Since then fourteen additional cases have been reported. These include a second recovery (in a nine months boy), which Mya reports in conjunction with other cases; and which Caccia (Archivio Italiano, 1903) fully describes. In this case also the lumbar puncture was employed; to be specific, three times with the evacuation of 35, 50 and 20 c.c. of purulent fluid. Mya, no doubt justly, ascribes the recovery more to the breast nourishment, which was continued throughout the disease, than to the lumbar puncture. In Mya's case of recovery there remained crossed paresis of the left facial nerve and of the right extremities.

It seems remarkable that several of the bacteriologically proved cases of influenza meningitis, exhibited other diseases, for instance otitis and pulmonary trouble, due to Pfeiffer's bacillus. In cultures obtained from these foci as well as from the meningeal exudate, which is constantly described as thick and purulent, there existed in some cases, besides influenza bacilli, diplo- and monococci and short delicate threads, which probably are to be regarded to some extent at least not as mixed infections but as division forms and involution forms of the influenza bacillus.

Finally it should be noted that Mya's three cases all occurred in the spring of 1902, when an epidemic of inflammations caused by Pfeiffer's bacillus prevailed in Florence. On the other hand, during the preceding decade, no analogous case was observed among numerous cases of meningitis which had been examined bacteriologically. This epidemic appearance certainly merits further study.

With the exception of a few microörganisms not well known and some of which have not yet been found in children (Stadelmann), we have now mentioned the chief topics in the bacterial etiology of diffuse purulent leptomeningitis.

From what has been said the **prognosis** also is apparent.

Treatment.—The therapy is as yet hopeless. Neither depleting by purgatives, nor reducing the blood supply of the skull, especially in the region of the mastoid process, nor counterirritation of the scalp by irritating ointments (inunctions with ung. tartari stibiati, P.G.), nor the local application of cold, cause tangible curative effects. The value of repeated lumbar punctures is likewise problematic, although *a priori* it seems rational to evacuate as much as possible of the pus and, by reducing intracranial pressure, to improve circulation and absorbtion. As the example of influenza meningitis has demonstrated, in many cases free employment of the puncture fails, while in one case (Langer) the withdrawal of a minimal amount of purulent fluid seems to have ushered in the improvement.

Major operations on the skull come into question only when primary pus foci in the vicinity of the meninges can be exposed and thereby constant or intermittent reinfection of the pia mater obviated. This holds good therefore especially in otogenous purulent meningitis.

When the diagnosis of purulent meningitis is established by all known methods, our efforts must be confined to the alleviation of the most distressing symptoms, which terrify the beholders. Chloral in large doses—0.5–1.0 Gm. (7–14 gr.) for an infant—in clysmas, and morphine appear to us better than baths, etc. Nothing remains to be done but to strive for euthanasia.

MENINGOCOCCUS MENINGITIS

(So-called epidemic cerebrospinal meningitis)

The propriety of describing this disease as a special form of purulent or seropurulent meningitis, a distinction which we have not made with regard to coli, influenza or any other purulent meningitis is primarily based upon clinical observation. If the typical cases, in which a careful bacteriological examination has been made, are selected, and all atypical cases, whose relation to this class is doubtful, are for the time being, excluded, it appears that we have to deal with a specific disease, of which the meningococcus intracellularis is the definite cause. Knowing this, we are then in a position to sift the atypical cases and to gather them into the confines of this disease.

This view, however, has not yet been generally accepted and is opposed to another, according to which etiologically different cases of primary meningitis, with a protracted and at times favorable course and with a tendency to appear epidemically, are considered together under the designation "sporadic and epidemic" cerebrospinal meningitis. The latter view is supported especially by A. Fränkel, von Leyden and Goldscheider and others: we, however, think it proper to follow the former, which is defended especially by Jäger, Heubner and others.

Etiology.—The disease germ to which we ascribe this important rôle, was discovered by Weichselbaum (1887) at six autopsies on cases of cerebrospinal meningitidis. To distinguish it from Fränkel's diplococcus pneumoniæ, he named it *diplococcus intracellularis meningitis.* The special diagnostic features were the following:—

1. The cocci occurred free in the purulent fluid of the diseased meninges, but preponderated within the pus cells. Here they were often found in considerable number and in a form strikingly resembling the gonococcus.

2. The cocci are always arranged in pairs, but in such a manner that the sides are juxtaposed (not the ends as is the case with diplococcus pneumoniæ). Often four lie together. Among the approximately even sized pairs, some decidedly larger ones appear.

3. The coccus grows only at body heat, best on agar, not well on blood serum, not on potato. The cultures form rather luxuriant, gray, viscid colonies.

These important observations of Weichselbaum attracted little attention (as in the case of Bordone-Uffreduzzi) and their validity was denied until in 1895, Jäger reported the same bacteriologic discovery in ten fatal cases in an epidemic which occurred in a barrack; and unreservedly affirmed Weichselbaum's diplococcus as the specific virus of epidemic cerebrospinal meningitis.

Heubner's researches caused a further important advance in this direction, since he first discovered intra vitam by lumbar puncture (in five children) the Weichselbaum meningococcus, which he designated intracellularis; and he succeeded by injections into the spinal canal of a goat in demonstrating its ability to cause a purulent meningitis.

Since then the Weichselbaum-Jäger-Heubner meningococcus has been the subject of numerous special investigations, partly concerning its clinical, partly its bacteriologic aspect, which we have no occasion to follow here. It should only be noted that exact study shows certain differences between the diplococcus described by Weichselbaum and the diplococcus which Heubner isolated and employed in his experiment on the goat. The chief of these is that the coccus of Weichselbaum is said always to be decolorized by, while Heubner's coccus retains, the Gram stain. Recently, however, Heubner demonstrated that the meningococci obtained from the same patient may behave differently toward the Gram stain at different stages of the disease, and that all other differences relating to the manner and luxuriance of growth, etc., which Albrecht and Ghon have emphasized, are inconstant or unimportant. In opposition to the effort to divide the meningococcus into two distinct types, viz.: Weichselbaum's and Jäger-Heubner's the view of Bordone-Uffreduzzi, until recently supported also by Concetti, Sorgente and others, may be briefly referred to, according to which the meningococcus only represents one variety out of the group of the pneumococci.

We believe we should omit a complete description of the cultivation and identification of the meningococcus, as this would be too long. Whoever wishes to study this matter will moreover be unable to omit studying the works mentioned and the original literature cited in them. Still it should be emphasized that the cocci are often limited in number and must be carefully sought in microscopical preparations. Nor is the cultural proof always easy, as these cocci do not grow at all on the common culture media, or at first they grow slowly and very delicately, transparent, and only gradually in progressive cultures show thick viscid colonies. In order to avoid the treacherous sources of error, it is advisable to follow the procedure of Heubner. With a sterilized pipette about 0.3 c.c. of the aspirated fluid is added to the water of condensation in an agar tube; after the tube has stood in the incubator 12 to 24 hours, the fluid is distributed over the surface of the agar by turning the tube. Then in 24–48 hours a rich growth occurs. From this first culture the propagation may then be carried on through many generations by simple inoculation.

The meningococcus intracellularis in many cases does not exist in pure culture, but mixed with other pyogenic cocci and bacteria, most frequently with Fränkel's pneumococcus. Besides this, pyogenic staphylococci and streptococci, sometimes influenza bacilli have also been found. Occasionally it occurs also in tuberculous meningitis as a secondary infection, in which case it seems not to influence the course of the disease.

It is always to be observed (which we desire briefly to remark), that there are various microörganisms (as, *e.g.*, the micrococcus catarrhalis, Pfeiffer), which microscopically appear identical with the meningococcus and resemble it so closely in cultures, that only the most accurate investigation prevents errors.

In cases of meningitis the meningococcus has been found sometimes alone, sometimes with virulent pyogenic bacteria, more or less numerousously present in most of the complications and also in the blood. In this connection the fact that it is frequently and richly present in the nasal secretion is most important, because its dissemination in the vicinity of the patient can only be from this source. Concerning its term of life and vital requirements outside of the human body the reports are so contradictory that no final decision can yet be pronounced.

Point of Entry.—The paths by which the meningococcus intracellularis enters the body are as yet not positively known. Based on the results of Weigert and Strümpell, who in their autopsies on cases of meningitis, had found pus in the pharynx and the adjoining cavities, the ethmoid plate was long considered the portal, and indeed the more certainly, since it has been established by sufficiently numerous examinations, that the meningococcus dwells on the nasal mucous membrane of sound persons and persons continuing to be sound who were in the vicinity of the patient. However this mode of infection is no more proven than the mode just assumed by Mastenhoffer—the third tonsil: moreover we know that such pyogenic processes may occur in all possible severe infectious diseases.

Many circumstances indicate an hæmatogenous infection of the meninges; however the source of the blood infection itself is still obscure.

The contagiousness of the disease is, as all experiences have shown, small in hospitals; but by no means to be underestimated, in unsanitary surroundings.

In barracks, prisons and in the dwellings of the poor, where cleanliness is often wanting, sometimes numerous contact infections occur. On the other hand it has been demonstrated by Peterson, that the infectious matter may also persist in infected sick rooms and thus cause the disease.

At any rate the fact that, even under the most favorable conditions, only relatively limited epidemics occur forces us to the conclusion that a few people only are susceptible to the disease. Why this is so is not known; often it is by no means the weakly children that are attacked. We can only point out a few predisposing causes, which temporarily increase the susceptibility of the individual. Continuous severe exposure to cold is occasionally mentioned; but it is more certain that even light cranial traumas (diving into water, Leyden: Rückenmarkskrankheiten, Berlin, 1874, Vol. 1), general concussions, sunstroke, physical and mental strain due to military marches, examinations, etc., are strikingly frequent in the history of the case.

Concerning the epidemiology of the meningococcus little can be reported, since the germ has been recognized too recently and it is justly questioned whether all epidemics described as cerebrospinal meningitis are attributable to it. Beginning with the period 1860–1870, greater or lesser epidemics of cerebrospinal meningitis have occurred periodically in various parts of Europe and North America, especially during the winter and spring. If these are distributed, as usually they were in recent years, very sparsely and in various districts of a metropolis, they of course appeared to individual observers to be sporadic cases, and not actually to be connected with an epidemic.

Children are most frequently attacked, and especially during the first year of life. The great mortality among the poorest classes has been repeatedly emphasized. It is not probable that poverty and unsanitary surroundings reduce the vital resistance, but certainly the conditions for dissemination are more favorable. Perhaps the germs are carried by domestic animals; in which purulent cerebrospinal meningitis has been demonstrated repeatedly, and also by vermin.

Pathology.—The pathologic changes caused by the meningococcus were first systematically studied by Klebs and later so thoroughly investigated by Strümpell, Albrecht and Ghon and others, that the subsequent investigations have made no important alterations or additions.

More frequently than in other forms of meningitis, an unequal distribution of the purulent exudate in the form of flakes and bands is found at various parts of the *cerebral cortex*,—especially over the parietal and occipital lobes and the cerebellum. The pia mater lying between the purulent areas is always slightly œdematous, and this œdematous fluid contains meningococci. If the pyogenic process involves the whole surface of the brain or, as not infrequently occurs, if it is markedly developed only at the base, the disease does not in any way differ macroscopically from meningitis of other etiology.

The intensity of the pyogenic process varies greatly if the cases run very rapid courses. After scarcely one day's sickness the purulent process may already be very decided, so that the entire surface of the brain is covered by a creamy, greenish yellow pus; but, on the other hand, the purulent process may be exceeded by the saturation and hyperæmia of the meninges, which as we have noted occurs not infrequently also in other fulminating forms of meningitis. Thus Sörensen reports a case, which terminated fatally on the fourth day of the attack, in which the autopsy showed only hyperæmia of the meninges, especially of the spinal dura. After a longer period of sickness, the exudate which at first was viscid, becomes stickier, firmer and may indeed have a consistence which calls to mind a thick slimy secretion. The flaky or bandlike distribution then appears very striking.

In the further course, if after weeks or months duration of the gradually recovering local disease, death occurs on account of complications or marasmus, one finds in the meshes of the pia only trifling remains of the amorphous exudate which has again become softer and more liquid.

The traces of the former pyogenic process may be recognized in local opacities and thickenings. But it is probable that gradually even these changes entirely disappear: at any rate we saw a case in the Breslau Children's Hospital in which, death having occurred from intercurrent disease months after recovery from a protracted meningococcus meningitis, the autopsy showed only an opaque area the size of a half dollar remaining on one parietal lobe.

If after the departure of the acute inflammatory process there remains pronounced and extensive thickening of the meninges, then gradual contractions of this scar tissue and obliteration of the important communications of the cerebrospinal canal are said to occur. Especially closure of the foramen of Magendi originating thus, is often described as the cause which leads to chronic hydrocephalus months after recovery from meningitis.

The changes in the *spinal meninges* are entirely analogous to the cerebral changes. In fresh cases thick accumulations of pus, especially on the posterior surface of the lumbar cord rarely fail. Also in protracted cases we find these in the form of flakes and bands at all possible parts of the spinal meninges. The participation of the spinal meninges in the pathologic and clinical pictures of meningococcus meningitis, regular and considerable as compared with other meningeal infections—has, as already remarked, led to its classification as cerebrospinal meningitis.

The chorioid plexus and the ventricular ependyma regularly participate in the meningeal affection. Less frequently encephalitic foci of hæmorrhagic-purulent nature are found. They may be situated as well in the cortex as in the medullary substance and through advanced disintegration cause multiple abscesses of the brain (Strümpell).

Characteristic changes of the internal organs do not occur or are caused by the extreme emaciation due to the protracted course.

On the other hand complications due to *metastatic pyogenic processes* occur not so infrequently. Of these articular and periarticular, sometimes also intermuscular purulent processes, purulent pleuritis, endo- and pericarditis, rarely dysentery-like enteritis, and nephritis may be mentioned. Of greater clinical importance than these, which often completely recover, are the disease of the eye and of the labyrinth, (which we shall later discuss more fully) since they heal—if ever--only with severe permanent defects.

Symptoms.—The clinical picture, the individual characteristics of which justify the classification of meningococcus meningitis as an independent disease, requires an accurate description. Heubner has so skilfully treated the points in differential diagnosis that we shall do well to repeat his words. Speaking of the sporadic cases, he says: "Their connection with the epidemic form is clinically characterized by the marked prominence of the motor and sensory symptoms of irritation; by the rigidity of the neck, spinal column, muscles, the violence of headache and of dorsal pain, the frequent recurrence of emesis, by the striking inferiority of the psychic disturbances, the clear intellect preserved throughout the greater part of the sickness, which renders the numerous painful experiences particularly tormenting and thereby creates extreme moodiness, by incessant variations in its course, general improvement lasting hours or days, which is always associated with striking change of temperament and is always dissipated by a fresh relapse, by the indefinite course, protracted for weeks or months, which however may still terminate in complete recovery, by the decidedly higher percentage of recoveries than appears in all other forms of meningitis."

Any one who has frequently had the opportunity of observing these usually sharply defined characteristics of epidemic meningitis, will hardly comprehend how these cases can be so indiscriminately classed with other forms of meningitis: the differentiation from some forms of tuberculous meningitis might perchance offer the greatest difficulty; but with purulent meningitis of other etiology only the fulminating epidemic cases can be confused.

As we have above described the clinical pictures of purulent and tuberculous meningitis and having here presented the characteristic features of typical cases of meningococcus meningitis in the quotation from Heubner, there remains the completing of the symptomatology at least in some respects, and the description of atypical courses.

We are indebted to Sörensen's researches for important conclusions:

The disease most frequently begins *suddenly*, indeed turbulently, with fever, sometimes with a chill, vomiting and pains especially in the head, less frequently in the limbs. Soon cloudiness of the intellect appears; the patient may be confused or delirious or stupefied; at times also isolated twitchings in certain muscle areas, or general convulsions occur.

Although this turbulent onset is the rule, there occur cases with a more gradual or intermittent beginning, without having on this account a better prognosis.

To facilitate a better understanding of the complicated course of the disease—characterized also by sudden changes of symptoms—Sörensen analyses it into what he calls its elementary parts, which "in their simplest form consist of fever and evidences of pain, to which symptoms very frequently vomiting and somnolence, at times symptoms of decidedly depressing character are added." If these individual elements, which can without distorting the facts, be gleaned from the history of the case, are separated by greater intervals more or less free from fever and pain, then there exists a plainly remittent character as well of fever as of the other symptoms. If on the contrary the elements directly succeed each other, then a more continuous course results.

Of course, besides these two types all imaginable variations occur in a disease so prone to changes and this also is exemplified by Sörensen.

In making a decision at the bedside it is important to know that, especially during the stage of invasion, the fever may not progress pari passu with the severity of the meningeal symptoms; but that with a rise of fever a diminution of the disturbances, previously very tormenting, may occur.

It must be regarded as a very unfavorable prognostic sign if neither exacerbations nor remissions of the fever induce noticeable changes in the general condition, and especially if the symptoms of exhaustion are not sometimes dissipated, at least during short periods of increased irritation.

Moreover in subacute fatal cases, the temperature before death may be normal or subnormal; usually, however, it exhibits in the last days a gradual rise persisting till death, such as we observe usually in purulent and not infrequently also in tuberculous meningitis.

The pulse, unlike that in tuberculous meningitis, is usually very rapid from the start, only retarded occasionally in intervals of apyrexia.

Taking into consideration the differential points previously described, together with Heubner's description, the clinical picture contains to a greater or less degree all the typical symptoms of meningitis. General epileptiform convulsions may, especially in young children, accompany the rise in temperature in the beginning of the disease; in its further course are observed clonic spasms involving a limb or one side of the body; and which are sometimes succeeded by paralysis of the part affected. Paraplegic paralyses without antecedent convulsions are no doubt usually of spinal origin (Strümpell). Clonic twitchings in the region of the facial, of the external ocular muscles (nystagmus) and of the hypoglossus occur not infrequently. In all of this there is no typical difference from the other meningitides.

The hypertony of the entire musculature appears especially as extreme cervical rigidity with board-like tension of the upper portion of the sternocleidomastoid muscle, often as trismus and grinding of the teeth. Correspondingly increased reflexes are rarely absent.

A very unpleasant hyperæsthesia and hyperalgesia occurs quite as constantly.

Every sudden glaring light, every loud noise, every forcible touch of the skin, every passive motion and change of position, usually evokes distinct signs of pain. During the attacks the face (notably the cheeks and conjunctivæ) is congested and the vasomotor excitability of the skin is increased (dermatography). Eating is occasionally inhibited by the excessive retraction of the head, and restricted to fluids and semi-solids, often however at least during the respites astonishingly increased. Emesis is frequent. When constipation exists it is no doubt partially the result of insufficient eating, partially of a contracted condition of the intestinal musculature, which is deducible from the presence of the boat-shaped abdomen.

The urine is, notwithstanding the high fever, usually light colored and abundant; small quantities of albumin and occasionally also of sugar have no diagnostic or therapeutic significance.

Very frequently there appears, between the third and sixth day of the disease, herpes labialis or nasalis: as yet meningococci have never been found in the vesicles. If the physician first sees the patient during an interval when the symptoms of irritation are not apparent, this herpes may assist in the diagnosis.

The observations of Göppert illustrate in an interesting manner the great diagnostic difficulties which may exist if the course of a whole day at least is not reviewed, but if conclusions must be drawn from a single observation. He found that only twenty of his forty-four cases showed cervical rigidity at isolated examinations, and indeed that only eight of twenty-three children less than a year old showed this which is counted the most constant symptom. On the other hand, besides fever and rapid pulse, the disease may present a complete clinical picture without "meningeal" apathy, vomiting, anorexia, etc.

Göppert distinguishes three types among the cases without cervical rigidity. In the first type intracranial pressure and expansion of the cranium dominate the clinical picture. The course resembles also in its malignity, simple purulent meningitis. "The second type of the cases without cervical rigidity comprises those in which tension of the fontanelles also fails us. The children, who have high fever, rapid pulse and hurried respiration, present not a single symptom except pain on passive motion, *e.g.*, when they are propped up, unless some rigidity may be perceived: there is not a trace of cervical rigidity. How easily this type may be overlooked is obvious." He distinguishes as the third type cases with waxy pallor and high fever; these may be mistaken for certain septic forms of purulent cystitis in infancy which they resemble. Here careful urinalysis is decisive.

The **duration** and **course** in meningococcus meningitis show much greater variations in single cases than in any other form, since the disease may terminate in a few hours or after many months (six to nine, Eichorst).

The very acute cases, which terminate fatally in less than twenty-four hours, are in general rarely recognized and only positively by a postmortem. As we have already remarked, extensive pyogenic processes must not be anticipated; capillary and venous hyperæmia of the leptomeninges and œdema of the pia with comparatively few meningococci constitute all the discernible pathology. These cases have been denominated *meningitis cerebrospinalis acutissima* or *siderans.* If the disease begins suddenly with the symptoms of cerebral hæmorrhage, that is, with loss of consciousness and hemiplegia or monoplegia, it is often designated *meningitis cerebrospinalis apoplectiformis.* This form does not always terminate fatally.

In contradistinction to the severe acute forms mentioned there are abortive cases, which, if they appear sporadically, are hardly diagnosible, at least in young children.

The great majority of cases run neither the turbulent course depicted nor the simple course. If the disease terminates fatally, this usually occurs at the end of the first or during the second week, often much later, after months, in a condition of extreme emaciation, either associated with the described complications or on account of general debility.

In these protracted cases the disease runs the intermittent or remittent course to which we have already referred. At the same time it is impossible to predict with any degree of certainty the probable outcome of the disease. Death may unexpectedly occur during a period of evident diminution of the cerebral symptoms.

The reported percentages of recoveries vary greatly from 25–30 per cent. (Florand) to 63 per cent. (Netter) and 68 per cent. (Kohts). Leyden and Goldscheider report from 20–70 per cent.

Whether all these cases really were due to meningococci is doubtful and must be determined by bacteriologic investigations. We believe that the high death rates indicate any meningitis occurring epidemically, such as may be caused by pneumococci, and must be separated from meningococcus meningitis.

It may be accepted as certain that the **prognosis** is much worse in the first and second years than later.

Complications. Meningococcus meningitis has more complications than the other meningitides. Apparently these originate metastatically, rarely or probably never by direct extension from the diseased meninges. These are diseases of the eye and ear as well as the secondary development of chronic hydrocephalus. Furthermore there occur, as already mentioned, articular and periarticular pyogenic processes, in the pleura, endo- and pericardium. First, with reference to the *ocular complications*, we refer to Heine's classification, which also embraces the most important extracts from Knies, Schmidt-Rimpler and others concerning the numerous injuries, which the motor and visual functions as well as the anatomical integrity of the visual apparatus may undergo. Let it be noted that visual disturbances rarely are of cortical origin, most frequently neuritic basilar, not infrequently, however, due to an inflammation of the interior of the eye itself. This consists in a metastatic iridocyclitis, which often appears on one side but may be double. Hypopion appears only with severe iritis, otherwise most frequently the optic lens becomes cloudy. It shows no pyogenic tendency but, usually by the development of a so-called pseudoglioma or amaurotic cat's eye leads to permanent loss of sight. It should be mentioned that this metastatic ophthalmia may occur also in lighter forms, which apparently may not induce pseudoglioma with resultant blindness. It is remarkable that these milder forms are not infrequently double, while pseudoglioma is almost invariably single.

This ophthalmia accompanies not only severer cases of meningitis: it occurs more frequently as the most important localization of the meningococcus in light, indeed in abortive cases whose obscure cerebral symptoms are first correctly interpreted through the eye-trouble.

In this respect there exists a striking and interesting analogy with the most important ear disease which occurs with meningococcus meningitis, acute otitis intima, or *labyrinthitis*. This disease also occurs in severe cases and belongs to the early symptoms, but it seems to preponderate in the lighter or lightest cases.

As evidence of this it may be mentioned that it was originally described by Voltolini as an independent disease beginning with indistinct cerebral symptoms and it was only recognized as an incident of cerebrospinal meningitis much later. Clinically it is characterized by its occurrence on both sides, by the presence of another labyrinth symptom—dizziness—and by the severity of the functional disorder.

With the disease of the labyrinth there may be associated *otitis media*, as in any meningitis or in any severe general disease, but even then the complete irreparable deafness after the disturbances of equilibrium have ceased, indicates that not the middle ear alone has suffered.

The third of the complications of meningococcus meningitis mentioned above, *chronic hydrocephalus*, naturally cannot be an early symptom, since its development requires weeks at least. Either the inflammatory process persists in a weak form for months or years (Ziemssen), or mechanical factors, which it has caused (obliteration of important passages), or both factors together keep the intracranial pressure constantly above normal and lead to continually increasing ventricular dilatation with its well known clinical results. Arrest and relative recovery gradually appear in many cases, but not infrequently hydrocephalus is the cause of "late death" after apparently complete recovery.

Diagnosis.—The diagnosis of meningococcus meningitis, which, in many cases may be so easy and safe, can only be made by lumbar puncture in atypical cases. Increased pressure of the fluid is not invariably present, but we always obtain a punctate, turbid and purulent or containing gross pus floccules, in which we almost invariably find polynuclear leucocytes and meningococci. It has already been mentioned that these latter may be scarce, to such a degree that a culture is indispensable for their positive identification. Hygienic institutes and bacteriological bureaus founded for the investigation of epidemics devote themselves to this necessity of medical practice.

In exceptional cases even the lumbar puncture may be indecisive, particularly under two conditions. Either the pus in the spinal canal may be so thickened that it cannot discharge through the cannula, in which case it can be diluted by injecting a small quantity of sterile salt solution; or the purulent process is limited to the cerebral meninges and by adhering to the occipital foramen is prevented from flowing into the spinal canal. Then in spite of the increased intracranial pressure a small quantity of more or less clear liquid is obtained, in which however meningococci can usually be found in cultures.

Treatment.—The treatment of meningococcus meningitis is still chiefly symptomatic. The hyperæsthesia of the patients requires a quiet, comfortable position and the greatest possible avoidance of all painful manipulations. For small children, on account of the excessive tenderness of the spinal column, a plaster of paris bed or a similar firm bandage has occasionally been employed with success. During the paroxysms of pain morphine in ample doses may be given without hesitation, if chloral, trional, phenacetin, antipyrin, etc., have failed.

Concerning the application of cold to the head, neck and along the spinal column it appears proper to proceed according to the sensation of the patient; *i.e.*, to relinquish this treatment if it affords no relief or is unpleasant to the patient. Also local blood letting has been both condemned and lauded.

That a disease leading to such severe emaciation, through difficult nutrition (opisthotonos), vomiting, persistent fever and probably specific trophic disturbances, demands the most careful nourishment and nursing requires no further discussion; but it should be remembered that fluid nourishment, which is most necessary, should not consist exclusively of milk, but should conform to the general principles of nutrition.

Two other therapeutic measures should be especially considered: first, baths; second, lumbar puncture.

On account of the excessive sensitiveness of the patient, cooling *baths* cannot as a rule be employed, and careful packs or similar procedure must be substituted for them. On the other hand warm, or rather hot baths, since their recommendation by Aufrecht, have established their right to a place in the treatment of cerebrospinal meningitis. They should be given at 40° C. (104° F.) and continued ten minutes or long after this. The skin should be flushed and should undergo a long sweat in an immediately succeeding pack.

Their mode of action has not yet been satisfactorily explained and their reckless employment for all patients is not justified. At least the first of such baths in every case should be given under the physician's observation of the pulse, respiration, etc. The same precaution is certainly requisite in the similarly dangerous pilocarpine treatment.

Lumbar puncture, to the diagnostic value of which we have already referred, has been frequently employed and recommended for diagnostic purposes. It is conceivable and experience confirms the fact that aspiration of an amount of fluid sufficiently large to secure normal pressure, may temporarily alleviate headache and other symptoms related to increased intracranial pressure. It is probable also that thereby the intracranial circulation may be made more nearly normal and more favorable for resorbtion of the exudate. Nevertheless it remains questionable whether this effect is not of much too transitory a nature to produce a tangible improvement, even if the puncture is repeated frequently, *i.e.*, at intervals of one or a few days. Clinical observation and statistics do not prove conclusively the permanent curative effect of this treatment or even the possibility of preventing a secondary hydrocephalus. On the other hand the operation may be regarded as so safe that it should always be employed as a palliative.

The unsatisfactory curative effects of simple lumbar puncture have led to supplementing it by irrigating the spinal canal with sterile normal salt solution or the injection of antiseptics. Both procedures have their advocates. Franca, after aspirating 25–30 c.c. of cerebrospinal fluid, injected 3–9 c.c. of a 1 per cent. lysol solution into the spinal canal and repeated this treatment sometimes daily. His good results have not been confirmed by other observers.

Likewise discouraging results have as yet been obtained by incising the dura after lumbar puncture—as Quincke already did—and by more radical operations, *e.g.*, trephining the skull or incising the membrana obturatoria posterior. These methods were tried exclusively in older patients.

It should be briefly noted that the internal medical treatment of cerebrospinal meningitis with sodium iodide and hexamethylenamine has as yet accomplished no results above criticism.

Flexner recommends the spinal injection of a bacteriolytic serum obtained from many strains of the meningococcus. The results show a larger percentage of recoveries than by any other treatment. Of 1500 cases treated with the serum in many countries 75 per cent. recovered.

SEROUS MENINGITIS

The term serous meningitis is primarily anatomical and as such is ambiguous. As already mentioned in the introduction, tuberculous meningitis is a definite type of the serous form, which for practical reasons we have discussed separately. Furthermore a second type has been pointed out which is caused by fulminating infections with pyogenic germs, if death from toxæmia occurs before the purulent process has had time to develop.

Another type is characterized by a more or less acutely beginning exudation of the meninges, perhaps of the ependyma, which is excited by any kind of inflammatory irritation, concerning the nature of which we shall speak later on, and which permanently or at least for a considerable period betrays no tendency to become purulent. In such cases the serous exudate is usually not copious and only distinguishable from the normal cerebrospinal fluid and fluid altered by stasis by microscopic chemical peculiarities. As also the histological changes of the meninges, of the ependyma and parts of the brain lying beneath them are not decidedly marked and are difficult to interpret, it is easily understood that for a long time there existed misconception of the relations between the trifling post-mortem disclosures and the oftentimes severe meningeal symptom-complex: and this gave rise to the designation "pseudomeningitis" or "meningismus."

Dupré, who coined the word "*meningismus*" defines it as follows: "the complex of symptoms caused by the lesions of the meningocortical zones and independent of any perceptible anatomic change." Thus he really discards all meningeal and cortico-encephalitic sources, however, as was later recognized, unjustly.

Some of the conditions which Dupré and many of his successors describe as meningismus, are without doubt cases of serous meningitis while others are conditions which are probably caused by autointoxications or vasomotor disturbances and which we are accustomed to count among functional disturbances so long as we do not understand the fine cellular pathology which causes them. The view, that the term meningismus embraces heterogeneous diseases and is convenient rather than scientific, very soon curbed the great enthusiasm which the word originally aroused, and to-day not only German but also nearly all the French authors advise its abandonment.*

Returning from this digression, to serous meningitis, it is possible when we have to deal with a case of meningitis to determine its nature primarily by employing lumbar puncture. The cerebrospinal fluid is seldom under increased pressure. The aspirated fluid is clear: but the fact that it contains cellular elements, fibrin, and that its albumin is increased is evidence that meningitis is present, and enables us at once to make an important differential diagnosis from the functional disorders due to intoxications or osmotic disturbances.

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* The word pseudomeningitis may be advantageously reserved for meningeal symptom-complexes of purely hysterical origin.

The rising intracranial pressure, which in the beginning is often absent, may be recognized—by the abnormally increased tension of the fontanelles only in infancy—as is the case in the other meningitides. In older children it can be established only by lumbar puncture.

The microscopical demonstration in the sediment obtained by centrifugation, of variable quantities of leucocytes, mostly small mononuclear; the formation of a delicate spiderweb-like fibrin coagulum; and the abnormally heavy precipitation of albumin by Brandenberg's albumin test establish the inflammatory origin of the disease, but alone do not enable us to differentiate between tuberculous and other forms of serous meningitis. This portentous decision can only be made by reviewing all the other clinical points which the case presents, and by bacteriologic investigation. Particularly under these circumstances the demonstration of tubercle bacilli, the technic and difficulties of which we have commented upon, attains great significance.

In every suspicious case the cerebrospinal fluid, obtained with aseptic precautions, should be studied culturally and by animal experiments. While a positive result of bacterial examination is of great value, conversely a negative result must be estimated most carefully, since it by no means proves the absence of all microörganisms. We shall return to this point.

Serous meningitis, no doubt very rarely occurs as a primary disease in a previously healthy child; usually it appears as a complication in the course of various infections. In infants pneumonia and gastroenteritis; in older children, whooping-cough, measles and purulent otitis media are most frequently the primary diseases.

Pneumo- and streptococci and bacterium coli seem to be the commonest infectious agents; but besides these typhoid bacilli, staphylococci, and influenza bacilli have been found. It is noteworthy that only small numbers of all these microörganisms are contained in the spinal fluid of serous meningitis; while in the purulent forms large numbers are easily found. According to the anatomic localization of the inflammatory process it is possible to distinguish an acute external and internal serous meningitis but of course they may appear together.

The macroscopical pathology in the *external form* consists chiefly in œdema of the leptomeninges which in its scope may involve both base and convexity or be confined to islands in single larger or smaller areas. The differentiation from congestion œdema of the pia is only possible by microscopic evidence of inflammatory infiltration of the meninges, particularly of the lymphatics of the small vessels of the pia and cortex. Concerning this numerous investigations have been published during the last twenty years.

In the internal form we find dilatation of the ventricles and flattening of the convolutions; the fluid is usually clear, with slight changes which indicate inflammation of the chorioid plexus or ependyma.

In acute cases hydrocephalus never attains very extensive development: when this exists moreover, it implies a chronic, perhaps a chronic intermittent disease, to the consideration of which we shall return.

The *clinical picture* of serous meningitis exhibits various types and courses within the limits of the meningeal conditions.

A fulminating form, "apoplexia serosa" of the older authors, is exhibited in infants by convulsions beginning suddenly with high fever, sometimes hyperpyrexia and coma.

Apart from slight cervical rigidity, which moreover is not always present, and contracted pupils, nothing points to meningitis. Death occurs within a few hours or day. Finkelstein has published some very instructive observations of this intricate condition which were cleared up by the autopsy.

At the same time he lays stress upon the fact, in which we thoroughly agree with him, that in such cases the severe convulsions do not depend upon a particularly virulent infection, but upon an abnormally violent reaction of the affected children, who invariably have given evidence of the spasmophile diathesis before this disease.

In less frequent, favorable cases the turbulent onset is succeeded by a clinical picture, which displays pronounced meningeal features and, as Finkelstein says, depends chiefly upon the external form of serous meningitis. Much more frequently this type does not begin so abruptly, but more gradually.

Ventricular serous meningitis, the so-called *acute hydrocephalus* of childhood, has been recognized much longer and is better understood.

"From the external form it is clinically distinguished by the accentuation of pressure symptoms. For practical purposes it may be grouped into two divisions, viz.: cases with acute onset and in which convulsions predominate and cases with insidious development in which coma predominates" (Finkelstein).

The *convulsions,* which usually occur during the course of a more or less severe infectious gastro-enteritis attended by fever, differ from simple eclamptic convulsions primarily in their longer duration.

While eclamptic attacks continue scarcely longer than a few minutes, and in the status eclampticus recur repeatedly at short intervals, the convulsions due to serous meningitis may last for hours or days with only isolated interruptions, exactly as is the rule in other organic cerebral affections. Clinically, the convulsions resemble those caused by autointoxication, circulatory or metabolic disturbances of early childhood, which Thiemich has described as terminal.

A further important differential point between functional convulsions of pyretic origin and those due to meningitis is the possibility of observing at least to some extent cervical and spinal rigidity, hypertony, contracted pupils, somnolence, etc., during the variable intermissions between and at the conclusion of convulsions.

In the insidious variety the resemblance to *tuberculous meningitis* may be so great, that only careful examination of the aspirated cerebrospinal fluid prevents errors, which otherwise would be unavoidable. This error probably has often caused the publication of cures of tuberculous meningitis, when the cases really were simple serous meningitis. The mistake is more readily made when the child which presents symptoms of meningitis, has some other tuberculous affections; although even such cases not exceptionally are only serous meningitis.

Acute inflammatory hydrocephalus attains especial clinical significance in that, if the child lives, it becomes chronic and then either exhibits the progressive course of the common chronic hydrocephalus and ends fatally, or becomes stationary after moderate development, but is still ominous for the afflicted children.

In the first place these comparative recoveries often occur after the mental or motor functions of the brain have already suffered and in the second place the existence of a limited chronic hydrocephalus is a constant danger, a point of lowered resistence for the remainder of life. To Quincke belongs the credit of having called especial attention in his studies of serous meningitis to those cases which on account of trauma, physical or mental overexertion or excesses, or infectious diseases before puberty have acute or subacute exacerbations of the exudative process, which may be a menace to life. Infections, which are the chief *primary* etiologic factor, are no doubt rarely the cause of these exacerbations; as Quincke says aseptic angioneurotic processes are more likely the cause. Indicative of this fact are the rather slow onset, the feverless course, the absence of bacteria in the aspirated cerebrospinal fluid, which always is evacuated at high pressure; and, not least important, the result of lumbar puncture is decidedly more favorable than is seen in other meningitides.

Finally of great theoretical interest is the fact now firmly established, that in a number of affections (gastro-enteritis, pneumonia, etc.), bacteria are demonstrable in the cerebrospinal fluid which may cause symptoms of cerebral irritation of more or less pronounced meningeal type merely by toxic action upon the cortical parenchyma without histologic changes of the meninges or of the superficial supportive structures. It is impossible to say how frequently this occurs or presents clinical symptoms.

All that is important concerning the diagnosis and prognosis of serous meningitis has now been touched upon.

Treatment.—Its therapy is more promising inasmuch as generally we have to deal with a secondary condition which, after the subsidence of the primary focus of infection, recovers. Therefore the therapy should primarily be directed toward the basic disease, gastro-enteritis, pneumonia, etc. Finkelstein places above all other considerations the necessity of careful examination and treatment of the ear, by which he often secured surprisingly rapid disappearance of the cerebral symptoms.

Favorable measures in the form of cold packs or hot baths (highly recommended in cerebrospinal meningitis), and which perhaps should be repeated several times daily, have good effect.

Inunctions of gray ointment are recommended especially by Quincke, because of his good experiences, and have been successfully employed by others. On the other hand Quincke's recommendation to cause a pyogenic process as a counterirritant by applying unguentum tartari stibiati (P.G.) to the scalp has met with little favor.

The most important therapeutic measure is without doubt lumbar puncture, which by diminishing pressure secures more favorable conditions for circulation and absorption. This especially is the case in the ventricular form.

THE MOST IMPORTANT DISEASES OF THE SKIN

(Except Tuberculous Skin Affections)

BY

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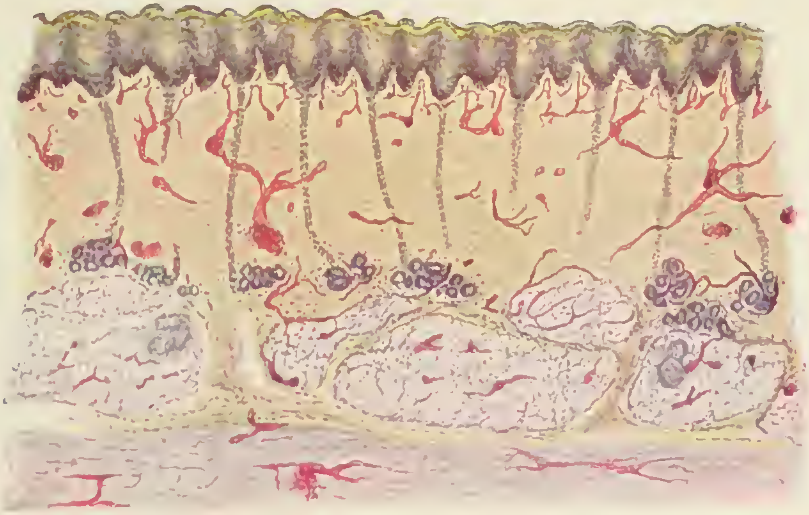
SURVEYING the diseases pertaining to the periods of infancy and early childhood, we are struck by the large number of skin affections, which far exceeds that of any other period of life. Scarcely an infant or young child entirely escapes some kind of skin affection, be it ever so slight: one suffers from intertrigo, another from eczema and another from urticarial manifestations. Very few children remain absolutely free from skin troubles. This is easily explained by the facts that the skin of the newborn has a special tendency for such disorders; that a large number of interior or exterior injuries exert their influence upon the skin of the newborn; that heredity and various diatheses contribute to the abnormal frequency of diseases of the skin.

The *skin of the newborn infant* is soft and tender, in which condition it remains during the first few years of life; the epithelium is disproportionately thin and the epithelial cells are less coherent than those of the adult. The corneal layer has very little power of resistance, and there is slight but regular desquamation. The papillary layer which is only slightly developed, has a very rich vascular plexus which in the first period of life is much more sensitive than later. The blood vessels therefore easily allow serum to exude, which may enter the connective tissue as in urticaria, or may lift the epithelium in the shape of small vesicles. The sweat glands are less strongly developed than the sebaceous glands, resulting in a very strong secretion of oil which finds expression in a great accumulation of seborrhœic masses in early childhood. Again, soon after birth a change of the hair takes place which has already been prepared during the last part of the fœtal life; and all these factors contribute to a predisposition for skin affections. It is especially the overaccumulation of oil which irritates the skin and is responsible for quite a series of dermatoses. After the first period of childhood the skin follows a regular course of development and only at the time of puberty is the skin again disturbed. We know that at this period, when the genitals develop, a marked growth of hair takes place and that the epithelial glands undergo decided development. Consequently there is aside from the growth of hair a considerable accumulation of corneal cells in the follicular infundibula, frequent inflammation in and around the follicles (appearance of comedones and acne pimples) and an increased reappearance of seborrhœic manifestations.

In the second place the body of an infant is exposed to *exterior and interior injuries.* We know that both the excretions and secretions of the skin may have an irritating effect. There is an overflow of saliva or, the milk runs down the nursing baby's face irritating the skin by decomposition, the feces and urine soil the child and through insufficient attention and cleanliness are the cause of many a dermatitis so frequent in the nursing period. On the other hand, *excess of cleanliness* acts just as injuriously upon the skin as does its absence through decomposition. Frequent washings and rubbings easily deprive the skin of its fat protection and remove the sebaceous secretions, exposing the dry and thin epidermis to all kinds of injurious exterior influences.

Fig. 65.



Skin of the newborn child. Epithelium thin, papillary layer slightly developed with rich vascular plexus. At this place the sweat-glands are well developed.

We know further how vigorously children scratch themselves and that, contrary to the habits of adults, give themselves up to the irritation of itching, thereby effecting the entrance of dirt into the skin by the action of their finger nails and fingers. A long series of impetiginous and furunculous diseases of early childhood are thereby explainable.

We also know that *toxic substances* may be resorbed from the intestinal canal into the blood, finding an entrance into the skin by way of the circulation. Not infrequently autointoxication produces disturbances of assimilation, forming a chief cause of skin affections from the intestine (urticarial affections, etc.).

In the third place *microörganisms* or their toxins may enter the circulation from the intestine and thereby cause a dermatosis. In this respect I need only remind of the still unsolved question of the origination of erythema exudativum through bacteria or their toxins.

In the fourth place it is a well-known fact that *nerve action* may produce reflexes which not infrequently produce dermatitis. As to the *influence of dentition* upon skin affections, there is as yet no conclusive proof of there being a mutual connection, but it is generally conceded that the period of dentition renders the skin more susceptible to pathological conditions.

Finally, it stands to reason that excessive or *inferior nourishment* exerts an unfavorable influence upon the skin. Badly nourished, atrophic or cachectic children are more susceptible to skin diseases than healthy ones who are powerful enough to resist them. On the other hand, *overnutrition*, especially during the nursing period, is responsible for excessive fat cushions in anæmic children, thereby favoring disorders of the circulation and with it the appearance of eczema, a very frequent complaint and generally recognized under the name of milk eczema. By changing the conditions of nutrition, the eruption may be materially improved if not entirely cured.

Beside the physiological and injurious conditions of the skin, *heredity*—the hereditary family taint—causes quite a number of skin diseases, or is responsible for a tendency thereto. It is generally known that ichthyosis and other keratoses are a family affection and that xeroderma pigmentosum and epidermolysis bullosa occur only in certain families. It is also a well-known fact that there is a special tendency for eczema in certain families, in some of which every member responds by an eczematous eruption to an external irritation.

There is a close connection between heredity and *diathesis* as to the influence exercised upon the skin. Diathesis also is hereditary, although its action on a child is different from that on the adult. A scrofulous and lymphatic (exudative) diathesis especially has an undoubted influence upon the skin. Whoever has had an opportunity of observing scrofulous children with their tendency to catarrh of the eyes, chronic coryza and enlargement of the glands, will know how frequently scrofulous eczema with its characteristic general features occurs among them, how the eczema frequently opens the door to the tubercle bacillus and thus to scrofuloderma and invasions of lupus, and how a chronic nasal catarrh frequently changes into lupus of the mucous membrane.

On the other hand, a lymphatic diathesis with its hyperplasia of the entire lymphatic apparatus, its enlargement of the spleen and lymphatic glands, hypertrophy of the palatal tonsils, and of the adenoid tissue in the nasopharyngeal space, prepares the soil for quite a series of skin affections.

The frequent occurrence of rheumatic affections in erythema multiforme and nodosum has been ascribed by the French (especially Bazin, Besnier and others) to a connection with arthritic diathesis.

HYGIENE OF THE SKIN

Hygiene of the newborn must of course be adapted to the above conditions. As this subject has been considered in another place, it will suffice at this juncture to state the most important hygienic measures for the first period of the child's life.

It goes without saying that the principal care of the skin consists in keeping it free from secretions and excretions, and to cleanse the skin from dust, sweat and fat. This is best accomplished by *washing* the child with warm or lukewarm water and mild, nearly neutral, nonirritating soap. Unna's "neutral soap" can be recommended as the best children's soap in all cases. Next to washing the child in point of importance is the *bath*. The average temperature of the full bath for the newborn should be 35° C. (95° F.) and the length of time five to ten minutes every day until the second year; at the end of the third year the temperature may be gradually reduced to 32–33° C. (89.3°–91.2° F.); from the third year the child need only be bathed three or four times a week, later twice a week. Certain substances may be added to the full bath, the mildest of which is wheat bran. One-half to three pounds placed in a linen bag are boiled for half an hour in about five litres (5 quarts) of water, the decoction being poured into the bath. This is rightly considered as having the mildest and most soothing effect upon the skin of the newborn. The next best baths are the astringent ones, prepared by the addition of wild thyme or chamomile, a handful of which is likewise boiled in five litres (5 quarts) of water which are added to the bath.

Cold full baths or cold douches are not advisable in the period of infancy. Cold sponging after warm baths only comes into question at a more advanced period. According to whether the skin is dry or fatty, hygienic measures should be taken to render it either more or less fatty. Thus, after each bath and also during the intervals the skin should be either powdered or anointed. For severe seborrhœa of the head frequent washing with lukewarm water (to which chamomile or wild thyme has been added) and soap, or with soda 3 to 5 Gm. (45–75 grains) soda to 500 c.c. (16 oz.) of water is to be recommended. To loosen the scaly masses the best and most convenient oil is weak salicylic oil on account of its keratolytic properties (salicyl 2–5 Gm. (30–75 grains) castor oil 40 Gm. (1 oz.) olive oil ad 100 Gm. (3½ oz.) to be applied after having been warmed in a water bath). With this oil it is possible to loosen all these deposits in a short time.

In conclusion I will briefly repeat what has already been stated at another place when treating of general therapy, that hygienic treatment also requires a sensible, sufficient and not overabundant nourishment, as both extremes in this regard can easily cause skin affections, as stated above.

TREATMENT

The object of the therapy of skin diseases consists in the first place in the treatment of the causative factors; in the second place in the alleviation of the subjective complaints; the curing of diseased portions of the skin; the protection of the skin from a spreading of the disease; the after-treatment; and prophylactic measures to prevent the return of the trouble. Therefore, the treatment will chiefly be external, and besides very often internal (medicinal or dietetic) if the pathological conditions may be assumed to have an internal causation. Prolonged after-treatment will be necessary, in order to protect the easily irritable skin from relapses. The treatment should be as simple as possible; the less medicine is given, the better, the dermatologist's duty consisting in the art of favorably influencing a disease without the promiscuous use of drugs.

Internal Treatment.—For the internal treatment of skin diseases comparatively few remedies are at our disposal. The one best known and best tested as to its efficacy is arsenic. The prescription for children is best given in the form of liq. potas. arsenicosi (Fowler's solution) 2.0 Gm. (30 ♏): aq. destill. 8.0 Gm. (2 drams) t.i.d. 5 drops (each 5 drops corresponding to 1 drop of Fowler's solution); instead of the distilled water it is often well to prescribe elixir aurantiorum compos. (P.G.), which the stomach bears very well. The dose for children from 1–2 years is 0.05 Gm. ($\frac{3}{4}$ grain) per dose; from 3–4 years also 0.05 Gm. ($\frac{3}{4}$ grain); from 5–10 years 0.1 Gm. ($1\frac{1}{2}$ grain); from 10–15 years 0.15 Gm. (2 grains). The effect of arsenic is best shown in psoriasis, acne, eczema and forms of prurigo and urticaria, although the effect is not certain and only slow.

Cacodylic acid and atoxyl have frequently been recommended as a substitute for arsenic. The former contains 54 per cent. of arsenic and its salt, cacodylate of soda; while the latter contains 37.69 per cent. of arsenic and is 40 per cent. less poisonous than Fowler's solution.

Pilocarpin is frequently administered subcutaneously or internally in all such pathological conditions in which it is intended to soften the skin without undue secretion of sweat (all forms of prurigo, etc.). The dose for injection up to the first year is 0.001 Gm. ($\frac{1}{60}$ gr.); from the first to the third year, 0.002 Gm. ($\frac{1}{30}$ gr.); from the third to the fifth year 0.003 Gm. ($\frac{1}{20}$ gr.); from the sixth to the tenth year 0.004–0.005 Gm. ($\frac{1}{15}-\frac{1}{12}$ gr.). Internally twice the subcutaneous dose is administered. Syrup of jaborandi can be recommended (and is always employed by me) as a substitute for pilocarpin. Children like to take it, and when given in tea, it produces almost the same sudoriparous effect as pilocarpin. Dose to three teaspoons.

Ergotin has been recommended by Henoch as a hæmostatic in all sanguineous exudations (purpura, etc.). Dose: Extr. sec. cornuti

0.25 Gm. (4 grains), aqua destill. ad 150.0 Gm. (5 oz.), (1 teaspoonful every 3 hours).

Lactic acid (colorless syrupy fluid) has been recommended by Du Castel, especially for all forms of prurigo which are referable to the intestinal tract. Dose, 3–20 drops a day in raspberry water.

Preparations of iron in conjunction with arsenic are very efficacious in the period of puberty, especially in anæmic and chlorotic children for the internal treatment of the sebaceous glands.

Ichthalbin (albumin of ichthyol; a fine grayish brown power, almost tasteless, is only resorbed in the intestine), acts as an appetizer with little children, regulates the peristalsis, also in dermatoses caused by engorgement (eczema of obese anæmic children) and in all intestinal disorders (urticarial conditions).

Oleum jecoris aselli (P.G.) (often superior even to phosphorated codliver oil) should always be given in lichen scrofulosum and in scrofulous eczema. It is recommended by Besnier especially also in prurigo. Dose: 4–8 teaspoonfuls, continued for a long time.

This practically exhausts the number of internal remedies which may be administered in childhood. It may be mentioned that weak doses of salicylic preparations may be prescribed in urticarial and erythematous conditions, and antipyrin preferably in conjunction with potassium bromide as a means to relieve itching in all pathological conditions in which there is violent pruritic irritation.

External Treatment.—Better success is achieved by external than internal treatment. In the first place baths are indicated to act favorably on the skin.

Aside from their general effect upon the body (acceleration of metabolism, circulation, etc.), warm or hot baths have a strongly macerating, keratolytic and detergent effect upon the epidermis and are therefore indicated in all hyperkeratotic processes, hyperæmia of the skin, urticarial and erythematous processes and the large number of eczematous eruptions as soon as the acute stage is over. To warm baths may be admixed with advantage: bran, chamomile or wild thyme, according to whether it is desired to produce a soothing or astringent effect. Baths with a more astringent effect are prepared by an addition of 2 to 4 pounds of oak bark (decoction) or walnut leaves (a decoction of a handful boiled in water). In France a favorite bath for children is made with starch, 1 to 2 pounds of which are dissolved in hot water. Of other baths there are the following:

Glue baths (1 to 8 pounds of carpenter's glue added to each bath), in frost-bites and prurigo.

Green soap baths in all pathological conditions where an increased keratolytic effect is desired, especially in psoriasis and ichthyosis.

Sulphur baths (wooden bath tub, not to be used in sitting rooms).

The sulphur bath may be given in three ways: By adding 20 to 50 Gm. (5–16 dr.) Vlemingkx's solution to the bath; by adding sodium hyposulphite (the weakest dose for children's baths being 50 Gm.); by adding 20 to 50 Gm. (5–16 dr.) sulphurated potash. Of natural salts for bathing purposes may be mentioned those of Aachen and Nenndorf.

Sublimate baths (wooden bath tub), 0.5 to 1 Gm. (7–15 gr.) per bath in all conditions where disinfection of the skin is intended.

Potassium permanganate bath, is less disinfecting than the former and slightly astringent (with the unpleasant by-effect of discoloring the skin). Dose: 3 Gm. (45 gr.) per bath.

Tar baths, to be prescribed for older children in all such cases in which a mild tar effect is desired (chronic, coarsely infiltrated eczema, psoriasis, etc.).

Salt baths.—These are prepared by adding 1 to 5 pounds of sea salt or Stassfurt salt to the bath, or 1 to 2 pounds of mother lye. For after-treatment of scrofulous eczema, but only after it has healed, also for long-continued treatment in urticarial affections. Caution should be exercised in all cases which have a tendency to eczematous dermatitis.

Aside from bathing purposes, water, in conjunction with various medicaments, is employed for moist bandages. Those most frequently used (with parchment or india rubber paper) are prepared with 3 per cent. boric acid, 1 to 3 per cent. acetic alumina, ¼ per cent. to 1 per cent. resorcin. Their effect is absorbing, desiccating and antiphlogistic in exuding eczema and bullous dermatitis. Weak sublimate bandages (0.1 per cent. solution) are employed for skin formation and healing of cauterized lupus foci.

Hot steam in the form of facial steam baths have been recommended, especially by Saalfeld, to remove comedones and oil in acne.

In conjunction with the bath treatment the employment of drugs is the principal part of the external treatment of skin diseases. This consists of powder treatment proper, treatment with fatty substances, ointment, pastes, paintable substances (glue, liniments, etc.). washing with spirits, plasters, and soaps.

POWDER

The effect of powder in the first place is that it forms a covering which protects against rubbing and external irritation; in the second place it dries and cools the skin and has therefore some antiphlogistic action.

There are vegetable and mineral powders.

Among the vegetable powders the following should be specially mentioned:—

Wheat flour (amylum tritici). This is unsuitable in exudative places on account of the tendency to form paste and turn sour.

Rice flour (amylum oryzæ). To be used on dry skin only.

Potato flour (amylum solani). The cheapest of the three flours.

Lycopodium seeds, yellow powder, a favorite in pediatric practice.

Among the mineral powders the following are serviceable:

Talcum venetum (finely powdered silicate of magnesia), soft fatty powder, often used in conjunction with drying powders (zinc, tannoform).

Magnesium carbonicum, frequently employed in intertrigo.

Vasenol powder, lately recommended.

OILS

We distinguish between animal, vegetable, and mineral oils, which are used partly alone or for the purpose of attenuating ointments.

(*a*) *Animal Oils.*—Codliver oil, to lubricate the skin, almost a specific in lichen scrofulosorum.

(*b*) *Vegetable Oils.*—Olive oil, and its substitution for practice among the poor, the cheaper rape oil (oleum rapæ); the finer and more expensive almond oil (oleum amygdal. dulc.); the thicker and stringy castor oil.

(*c*) *Mineral Oils.*— Petroleum. Slight antiparasitic effect (for frost-bites and as an antipediculosum); Paraffin, sterile transparent oil for the manufacture of white vaselin and stable ointments which will not get rancid.

FATS

White vaselin manufactured from the residue of petroleum. The American yellow vaselin is more uniform and reliable than white vaselin.

Nafalan and naphtalan (obtained from crude naphtha), especially in eczema, psoriasis, impetigo and prurigo and similar affections.

Lanolin (wool fat obtained from sheep's wool) does not get rancid, is an excellent constituent of ointments, absorbing water to over 100 per cent. of its weight. Adeps lanæ, at least equivalent to lanolin, absorbing up to 300 per cent. of water, recommended especially for the manufacture of hydrous cooling ointments. Type of cooling ointment: Adeps lanæ anhydric. 10 Gm. (2½ dr.) adeps benz. 20 Gm. (5 dr.), aqua rosæ, 40 Gm. (1 oz.). In conjunction with fats with 5–10 per cent. olive oil, 20–25 per cent. pig's fat to produce a good ointment mass. The cheap unrefined wool fat (oesypus) is used as a substitute for the expensive lanolin and adeps lanæ.

OINTMENTS

Every ointment should be of butterlike consistency, must admit of being finely distributed, and remain solid at body temperature.

Ointments chiefly used are: Adeps suillus (pig's fat), mixed with benzœ to prevent its getting rancid (adeps benzoatus).

Ung. diachylon Hebra (diachylon ointment: 5 parts lead plaster, 5 parts olive oil); the new prescription is better, ung. vaselin plumbic, because more stable and constant.

Ung. leniens (cold cream), a well known, mild, ointment base.

Ung. simplex (wax ointment), made from olive oil and wax. The latter two ointments are used to manufacture Neisser's cheap eczema ointment: zinc 1.0 Gm. (15 gr.), bismuth 1.0 Gm. (15 gr.), ung. leniens, ung. simplex aa 10.0 Gm. (2½ drams).

PASTES

A paste is a viscous, porous, desiccating salve, which contains equal parts of solid and fatty substances. It becomes harder at body temperature and is used as a protecting salve or bandage.

The following is a base paste of the simplest form and cheapest manufacture: Starch, vaselin q.s. aa m.f. pasta. The following base paste is a little more expensive but better, because it has somewhat better drying qualities: Zincum oxyd., amylum pur., vaselin, lanolin aa m.f. pasta. To this paste is added sulphur, tumenol, ichthyol, tar, etc., according to the effect it is desired to obtain, whether for instance desiccating and covering, as in alleviating pruritus, etc. The principal point in all pastes is that the percentage of the dry constituents is equal to that of the fats. The well known and very good paste of Lassar consists of acid. salicyl. 2.0 Gm., zinc. oxyd., starch aa 24.0 Gm., vaselin 50.0 Gm. Should it be desired to make these pastes softer, and reduce their porous and desiccating effect, and at the same time increase the softening effect, an addition of olive oil or paraffin oil will effect the purpose; for instance: Zinc oxyd., starch, lanolin, vaselin, ol. oliv. aa 10.0. The last-named paste can be had still softer by simply prescribing zinc and olive oil aa 50.0 m.f. pasta mollis (zinc oil). This zinc oil, introduced by Lassar, is one of the mildest and most protective prescriptions for infantile eczema.

GLUES

According to Unna the following two prescriptions make better protective coverings than pastes:—

1. Gelatina zinci mollis		
Zinc. oxyd. alb.	15.0	ʒiv
Glycerin	25.0	ʒvi
Aq. destill	45.0	℥ii
2. Gelatina zinci dura		
Gelatina	30 0	℥i
Glycerin	30.0	℥i
Aq. destill	30.0	℥i

In more recent times Unna recommended soft glues with an addition of 5 per cent. of gelatin and water, hard glues with an addition of 10 per cent. or 20 per cent. of gelatin without water. The most frequently used glues are those manufactured by Beiersdorf in Hamburg: Zinc glue, ichthyol glue, salicylic glue. The glue is heated in a porcelain dish or cup placed in a water bath, and then applied with a brush to the affected skin. Small pieces of cotton wool can be rapidly placed over the spot before the glue cools, after which a mull bandage is applied over it. The best results are obtained by glue in all dry, itching skin diseases of children who have to be restrained from scratching; it also saves the constant renewal of the bandages. Glues are of course contraindicated in exudative eczema.

WASHING WITH SPIRITS

Washing with spirits in itching skin affections, especially urticaria and prurigo, has a cooling and itch-relieving effect. Application: ¼ to ½ per cent. spirits of thymol, 1 per cent. to 3 per cent. spirits of carbolic acid, spirits of menthol with 10 per cent. of glycerin; with subsequent powdering.

DRY DUSTING

In itching, nonexudating, inflammatory dermatosis dry dusting has come into favor of late for the final healing of itching eczema.

Neisser and Boeck have furnished the following prescriptions:

Zinc oxyd.
Glycerin
Talcum.. ... ... .aa. .15.0. .ʒiv
Spiritus. aq. dest . .aa. . 7.5. .ʒi
(Neisser) m. f. liniment

Talcum powder
Amylum . . . . .aa. .25.0. .ʒvi
Glycerin . . . . .10.0. .ʒiss
Aqua plumb. . . . . .50.0. .ʒiss
(Boeck)

Linimentum exsiccans (Pick) and Unna's varnish ("Gelanthum") produce a thin pellicle to cover the affected places.

PLASTERS

Plasters represent a skin medication the consistency of which lies between that of salve and wax. The best plasters for pediatric purposes are either soap plasters or Unna's guttapercha plastermull ("paraplaste").

Among the best softening media for all chronic skin affections (eczema, psoriasis, etc.), and for all anomalies of cornification in which a very considerable softening of the corneal masses is required, as in corns, callosities, etc., are the following:—

Soap plaster with an addition of 5 to 20 per cent. of salicyl spread on a piece of linen according to Pick's instructions: Soap plaster with an addition of 2½ per cent. to 10 per cent. of salicyl spread on very soft "tricot."

The Beiersdorf plaster consists of a layer of caoutchouc plaster mass which is glued upon a thin layer of gutta percha. The other side of the gutta percha is covered with mull. The caoutchouc plaster mass may be mixed with 50 per cent. to 70 per cent. of medicaments. The plasters are soft and pliable, the medicaments are accurately dispensed, and in consequence of the impermeable gutta percha the plaster acts like a softening salve bandage under rubber paper. The most important plaster mulls for pediatric purposes are the following: Mild nonirritating oxide of zinc plaster (as adhesive plaster), chrysarobin plaster mull (in chronic eczema, psoriasis), mercury plaster mull and mercurial carbolic plaster mull (furunculosis, etc.), pyrogallus plaster mull (lupus), salicylic acid plaster (keratosis) also in conjunction with soap.

The great liability to tear and the dark color of the gutta percha mull have caused Unna to introduce the less destructible and almost flesh-colored "Paraplaste."

SOAPS

Under soap we understand the combination of fatty acids with alkalies. There are sodium soaps (hard) and potassium soaps (soft). Soaps cause a softening of the epidermis, desquamation and removal of corneal masses, also a swelling and loosening of the epithelium. The object of their use is that the medication may penetrate better into the skin after the removal of the corneal masses (especially in connection with baths). The application of soap is especially indicated for a skin abundant in fat which it is desired to remove. The effect may be increased if instead of simply washing with soap, the foam is kept on the skin, say over night, or even more by tying a bandage over the foam. In order to eliminate the irritation produced by the alkali upon the skin, attempts have been made to manufacture a neutral soap by overfattening, so that it should not contain either free alkali or glycerin. Unna and Eichhof who are deserving of great credit for their efforts in the direction of composing a medicinal soap, have introduced the following soaps, the applications of which I would recommend: Sulphur-resorcin salicylic soap (acne, comedones), sulphur-tar soap (seborrhœa, chronic eczema), sulphur-naphtol soap (prurigo and parasitic eczema), sublimate soap (disinfection), balsam of Peru soap (scabies), and tar soap (in all chronic inflammatory diseases of the skin). Soaps made of natural spring salts are the Nenndorf soap (16 per cent. to 35 per cent. sulphur soap), the Aachen sulphur soap, the "Krankenheiler" potassium iodine soap and the Kreuznach iodine bromide soap, all of which can be recommended in inflammation of the sebaceous glands. Whenever soap is used on a child, especially the medicated soaps, great caution should be exercised, as the infant's skin is exceedingly sensitive to soap treatment.

MEDICATION WITH SPECIFIC ACTION UPON THE SKIN

Among the medicaments which should be at the disposal of the pediatrist as well as the general practitioner in order to obtain the best results when mixed with salves, plasters, soaps, etc., I wish to mention the following as the most important and most reliable.

1. *Zinc oxide.*—Effect: desiccating, hygroscopic, nonirritating, antiphlogistic, indicated in all exudating skin catarrhs as a mild, desiccating addition to salves and pastes.

2. *Boric acid.*—Effect: weak, nonirritating, antiphlogistic and disinfecting astringent.

3. *Subnitrate of bismuth.*—Desiccating, acting as a cover medium, desquamative, as a mild addition to salves with zinc and boron.

4. *Tannic acid.*—Effect: hardening, vasculo-astringent, indicated in erythema, after-treatment for hardening eczematous skin.

5. *Salicylic acid.*—Effect: keratoplastic in small doses, keratolytic in large doses. Applied with salves, especially soaps and plasters.

6. *Resorcin.*—Strong medium of reduction. Effects desquamation of the superficial epithelial layer, at the same time blood reducing. Applied as a blanching and desquamative medium as an addition to paste in all affections of the sebaceous glands of the skin (in conjunction with sulphur) and in seborrhœa of the hairy part of the head.

7. *Pyrogallic acid.*—Strongest medium of reduction in consequence of its great capacity for reduction; very softening in old infiltrated and lupus areas. Acts as a specific in psoriasis. (Discoloration of light-colored hair, caution on account of danger of intoxication.)

8. *Lenigallol.*—Milder and less intoxicating than pyrogallic acid, lighter medium of reduction, acts excellently as an addition to zinc paste in slightly exudating facial eczema.

9. *Sulphur.*—Either as sulphur precipitatum, sublimatum or depuratum. Mild medium of reduction, effects loosening and swelling of corneal layers and desquamations; specific effect in folliculitis and furunculosis, affections of the sebaceous glands, etc. For baths, soaps, salves, etc.

10. *Ichthyol.*—Reduces congestion, is vasculo-astringent, antiphlogistic, administered both internally and externally. Indicated in all vascular dilatations, erythema, erysipelas, etc.

11. *Thigenol.*—Latterly recommended as a substitute for ichthyol, less odorous.

12. *Tumenol.*—On the market under the names of Tumenolum ammoniatum, T. pulverisatum and T. venale. Relieves itching very promptly, desiccating, keratoplastic, also heals eczema being practically nonirritating. Application: as tumenol zinc paste (Neisser) 5 per cent. to 20 per cent.; or as tumenol painting: tumenol 10.0 Gm., diluted spirits of ether, glycerin aa 30.0 Gm.

13. *Naphthol.*—Antiparasitic, promotes the resorption of infiltrates, relieves itching. Excellent desquamative medium (as desquamation paste), energetic effect in scabies and prurigo.

14. *Tar Preparations.*—In acute and subacute dermatosis, contracting the vessels, antiphlogistic, relieves itching, antiseptic. The following tar preparations are in use:—

(*a*) Pix liquida (wood tar), as solution with spirits for painting.

(*b*) Oleum rusci (birch tar).

(*c*) Oleum cadini (Spanish tar of cedar oil and juniper).

(*d*) Oleum fagi (beech tar).

(*e*) The last three as tinctures to be added to salves and pastes, for tar baths. Substitute frequently employed latterly: Lianthral (Beiersdorf's refined coal tar, less irritating than ordinary tar).

(*f*) Liquor carbonis detergens (light-colored spirit extract of tar) especially for application in the face and at the hands as an addition to white salves, for instance precipitate salves in psoriasis.

(*g*) Anthrasol (fluid, colorless tar oil). Mild effect, as an addition to pastes or salves or for painting if dissolved in one to thirty per cent. alcohol.

(*h*) Empyroform (almost odorless preparation of tar formalin). Mild tar preparation for paste and salve.

All tar preparations promote the development of comedones and acne, and should therefore not be used in these affections; they should also be avoided in exudative eczema on account of their irritating effect.

15. *Chrysarobin,* excellent preparation with energetic effect, keratoplastic, slightly astringent on the blood vessels, highly antiparasitic. Application: in all parasitic skin diseases, almost a specific in psoriasis. In applying chrysarobin (for children a weak ½ to 1 per cent. solution is best) great caution is necessary, as it easily causes considerable dermatitis with œdema and formation of vesicles, folliculitis and conjunctivitis and colors the hair dark (care about face). Should it create irritation, it should be immediately replaced by mild salves. Prescription: ½ to 10 per cent. salve and paste, 1 to 10 per cent. solution in chloroform for painting. Eurobin which is less irritating has recently been recommended as a substitute (application same as chrysarobin.)

16. *Formalin.*—Desiccating, antisecretory, antiparasitic. Application: as soap, spirits and salve.

17. *Menthol, thymol, carbolic acid* are used as strongly refrigerating, itch-relieving media in all forms of urticaria, prurigo and eczema, the best way being in the shape of salves and spirits.

18. *Styrax* (cheaper than balsam of Peru).—For vegetable and animal parasites (especially scabies); Peruol with castor oil (1:3) has of late been employed as a substitute for the highly odoriferous balsam of Peru.

19. *Hydrargyrum præcipitatum album.*—Effect: antiparasitic, blood-reducing and softening; excellent remedy in psoriasis and for destroying animal parasites (pediculi).

20. *Hydrogen peroxide,* with strong bleaching power like sublimate and oxychlorate of bismuth.

The last four medicaments are used in the shape of ointments for the removal of pigment and the black spots of comedones. Best method of application: Hydrogen peroxide 20.0 Gm. (6 dr.), adeps lanæ anhyd. 5.0 Gm. (1¼), vaselin 10.0 Gm. (2½ gr.), sublimate 0.05–0.1 Gm. (½–1½ gr.), bism. oxychlorat. 0.5–3.0 Gm.

21. *Nitrate of silver*, for painting in exudating eczema and intertrigo (2–10 per cent.). To clean ulcers as a one per cent. ointment with 10 per cent. Peru balsam. Instead of the nitrate good service has been rendered by Protargol-Vaseline (0.1: 10), which is nonodorous and nonstaining.

22. *Tuberculin*, as a diagnostic and therapeutic agent in tuberculous affections of the skin, especially of lichen scrophulosorum.

METHODS OF PHYSICAL CURES

The constant current is frequently employed for electrolysis (warts, small neoplasms, dilatation of vessels).

Electric light acts principally by the ultraviolet rays in the application of the Finsen or Finsen-Reyn lamp (lupus vulgaris). If instead of carbon light, iron light is employed (dermo lamp) the deeper structures will not be affected.

Röntgen rays relieve itching, dissolve old infiltrations, destroy neoplasms (in itching chronic eczema, warts, lupus vulgaris, depilation). On account of their serious by-effects to be used with caution.

Radium heals similarly to Röntgen rays; to be used with still greater caution.

ANÆMIA

Anæmia of the skin does not play any important part in children. Special mention should be made of the so-called marble-effect produced at the extremities, especially at the hands, by the effect of cold. It consists of small anæmic white spots which show distinctly against the surrounding bluish skin.

ERYTHEMA

Under erythema we understand a red spot caused by local hyperæmia. According to whether this erythema is an arterial or venous one, we have to distinguish between the arterial or congestive hyperæmia, and venous engorgement.

ARTERIAL HYPERÆMIA

The arterial hyperæmia of congestion is caused by an increased blood supply to the arteries, whether by augmented pressure or diminished resistance of the circulation. The best known form of this hyperæmia is *erythema pudoris*, produced under the influence of shame, joy, etc. It occurs chiefly in children and disappears with advancing age.

Soon after birth infants exhibit *erythema neonatorum*. It increases up to the middle of the first week of life and generally disappears without leaving a trace. Whether there is any connection between this affection of the vessels and the icterus which sometimes appears in its wake, is an open question.

In digestive disorders and in the period of dentition the so-called *infantile erythema* makes its appearance, sometimes in spots, sometimes spreading diffusely over the body; it is frequently accompanied by light fever.

In 1899, Sticker and Schmidt described an *erythema infectiosum* occurring epidemically in children, commencing symmetrically on the cheeks and spreading first to the extremities and then to the trunk. There are flat pimples, sometimes red and sligthly raised, burning spots, forming figures like geographical charts, and healing from the centre. They are often accompanied by dysphagia, coryza and light fever. The affection heals spontaneously in four to five days and is to be regarded as absolutely benign. It has also been described in 1904 by Placht under the same name (or megalerythema) as being a small family epidemic. Escherich classifies the affection among roseola.

To this category also belongs *erythema vaccinicum* (roseola vaccinica) and *erythema variolosum* (described in detail at another place). The former appears either on the second day or only after 7 or 8 days (and later still) after vaccination, either as a by-effect of the vaccine or poison or later in consequence of resorption of the ulcerous vaccine masses. I have personally seen such a case develop fourteen days after vaccination.

Erythema variolosum generally appears on the second or third day of illness.

The other forms designated as erythema, especially erythema from the effects of heat or counterirritants, erythema from the effects of light, are no actual erythemata, but only the prodromal stages of a dermatitis, the actual inflammation of the skin. When in the treatment of erythema the irritation is absent, the affection will not advance beyond erythema and the dermatitis will not appear. The best known is *erythema calorum* (sun burn or glacier burn), which is caused principally by the influence of the ultra-violet rays of the spectrum; among the forms of erythema from counterirritants there is chiefly erythema following the use of arnica, mustard plasters, etc. Under treatment by the X-rays, erythema and superficial dermatitis develop which under certain conditions may continue for a long time.

VENOUS HYPERÆMIA

Venous hyperæmia (engorgement) is characterized by the excessive dilatation of the veins of the skin and the engorgement of venous blood occasioned by a larger supply of blood than can be carried off. These engorgements are caused by anomalies of the circulation (cardiac insufficiency, etc., pressure of tumors upon veins, etc.), and the atony of the vessels, *i.e.*, the veins, following the preceding hyperæmia.

FROST-BITES

The first signs of freezing in infants as in adults are shown at the extremities (hands, dorsal surface of the feet, extensor surface of the fingers and toes) and at the peripheral portions of the face (nose, ears). Red or bluish red swellings of a doughy, œdematous nature appear which itch considerably and burn still more. They are popularly known as chilblains (perniones). After they have existed for some time, they become harder and more painful, vesicles form on them, or the surface undergoes ulcerous degeneration. If the cold is excessive or of long duration, the second stage of congelation immediately appears. Vesicles develop which are transformed into badly healing deep ulcers. The third and most severe degree of congelation sets in if a gangrene or necrosis of the deeper tissues spreads to the bones. The frozen part dies off with demarcated inflammation. In this way entire phalanges or large portions of the extremities may perish.

Anatomy.—Under the influence of great cold there is always an injury to the venous tonus which results in anæmia. As the effect of the cold ceases, the anæmia is followed by hyperæmia. The blood flows forcibly into the atonic vessels and a localized venous engorgement remains which is shown by its coloration. If the freezing continues the hyperæmia increases and finally leads to a continuously increasing transudation into the tissue. In the second and third stages of congelation hyaline and leucocyte thrombi arise (Hodara) and finally there is the picture of gangrene (Recklinghausen).

Pathogenesis.—The pathological condition arises also in healthy children through the influence of abnormally low temperatures or long-continued cold, but in weakly, anæmic children it takes place much more easily than in the healthy. In children it nearly always occurs in the shape of chilblains, and unless there is sufficient attention paid to it, it becomes more difficult to handle from year to year. This condition should be energetically treated from the beginning and the general constitution constantly improved, as otherwise the affection returns every year.

Prognosis.—The prognosis of congelation is therefore not very favorable on account of its great tendency to relapses. The chilblains may recur in midsummer, in July and August in damp, cool weather, and it requires the unflagging energy of the attending physician and the patient's family if the condition is not to become one of long duration.

Treatment.—The object of the therapy is in the first place to restore the lost tone of the vessels; in the second place to remove the affected parts by local treatment; thirdly, to improve the general condition to such an extent that the possibility of renewed attacks is diminished, and fourthly, to take prophylactic measures to keep injurious influences at bay.

1. Hot baths are most beneficial in order to influence the tone of the vessels. Hot water baths may be applied in the shape of hand and foot baths; or hot sand baths or medicated hot baths may be employed. The latter are prepared by adding decoctions of oak bark or walnut leaves (from a few handfuls boiled in hot water), alum, vinegar or glue. An exceedingly good effect in improving the tone of the vessels is obtained by changing hand and foot baths from hot to cold and vice versa (allowing the feet to remain a few minutes in the hot water and a somewhat shorter time in the cold water, the whole procedure to be repeated for a quarter to half an hour). This should be done every evening before the actual local treatment is commenced. In the morning I always prescribe rubbing with spirits of camphor in order to exert an influence upon the circulation. Electric baths have also been recommended in recent times, one pole to be applied to the body and the other to the affected hand or foot. The hands and feet should always be kept bandaged, at night perhaps provided with gloves and stockings. During the day the hands should be protected by comfortable soft woolen gloves, the wrists by warming cuffs, and the feet by well fitting stockings which should be frequently changed during the day on account of the perspiration. Narrow shoes are forbidden in order not to interfere with the circulation and to avoid pressure on sensitive places.

2. The local treatment depends upon the degree of freezing. Should there be chilblains with uninjured surface, where the skin is apparently intact, painting with 10 per cent. ichthyol oil or 10 per cent. Peru balsam, is advisable. If a salve bandage is applied, we use zinc paste with the following salve: 10 per cent. ichthyol, 10 per cent. Peru balsam and 10 per cent. camphor; or ichthyol 5.0 Gm., Peru balsam 3.0 Gm., camphor 0.3 Gm., naphtalan ad 50.0 Gm., which is very efficacious. The well known salve of nitrate of silver 1.0 (15 gr.), balsam of Peru 2.0 Gm. (30 gr.), ung. zinc. ad 20.0 Gm. (1 oz.) is also to be recommended. The treatment is best arranged so that the painting is applied in the morning, while the salve bandage is kept on over night, a foot bath having been given before applying it.

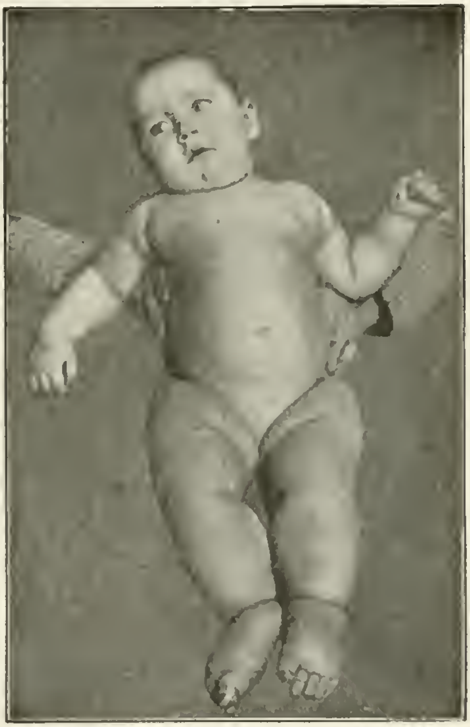
Open, eroded or ulcerative chilblains demand an irritating ointment which stimulates granulation. Camphor (10 per cent.), airolvaseline (10 per cent.), protargol, or nitrate of silver (10 per cent.) ointments are beneficial.

3. The treatment of the general condition consists in prescribing for the anæmia and ordering an invigorating diet. The following internal medication is recommended: Syrup of the iodide of iron, Iodide-ferratose (Böhringer), and all iron preparations. Fellow's syrup of hypophos-

phites and its German substitutes, the well-known tinctures of iron, arsenic, quinine, also strychine pills. Ichthyol capsules [twice daily 0.2 Gm. (3 gr.)] or its substitute Ichthalbin. The medicinal treatment also includes dietetic curative procedures (massage, hydrotherapy) and body exercises which are apt to stimulate the circulation.

4. By way of prophylaxis special care should be taken to avoid exposure to cold (cold floor in dwellings, prolonged skating, simultaneous exposure to wet and cold). During the summer, especially in the first years following the illness systematic treatment by baths and ointments placed over the affected parts should be instituted whenever there is a slight change in temperature.

Fig. 66.



Posterysipelatous œdema. Roselike sclerema following erysipelas).

ŒDEMA

If there has been considerable congestion for a long time, there will be exudation into the surrounding tissue. The skin becomes doughy, smooth and tense; pressure with the finger leaves an impression. Acute circumscript œdema as a variety of urticaria, appears suddenly and disappears just as suddenly. Myxœdema and elephantiasis (chronic œdematous elephantiasis), the later stages of which consist chiefly in connective tissue proliferations, will be dealt with at a later stage. In rare cases the so-called posterysipelatous œdema attacks children under one year in the wake of erysipelas (Henoch, Clementowsky, Von Holten). It is an inflammatory œdema accompanied by fever.

POLYMORPHOUS ERYTHEMA OF TOXIC NATURE

These forms of erythema are distinguished from those previously described by their multifariousness. There are really erythematous as well as inflammatory manifestations, so that they form the transition to the real skin inflammations. There are the following subdivisions: (1) erythema multiforme; (2) erythema nodosum; (3) the true polymorphous toxic erythemata; (4) medicinal exanthem; (5) serum exanthem.

ERYTHEMA EXUDATIVUM MULTIFORME (*Hebra*)

The pathologic picture first described by Hebra under the name of erythema exudativum multiforme is mentioned by other authors under names which are partly still in force to-day: erythema polymorphe (Kaposi), erythema hydroa (Bazin), erythema centrifugum, erythema marginatum, etc.

The general *clinical picture* is usually typical and uniform, appearing first at the dorsal surface of the hands and feet, then at the lower arm and leg in the shape of single raised spots which are at first the size of a lentil and become gradually larger. Their color is vivid, light red to brick red. After twenty-four hours at the most the centre of these spots sinks in and becomes cyanotic, while the vivid red remains only at the periphery in the shape of a circle. Gradually the number of these spots, or circles, increases (erythema iris), two or more of these circles run into each other, and the result is figures which are designated as erythema gyratum and erythema annulare. Beside these there may be occasional pimples or nodules resembling urticaria, or there may be raised spots resembling vesicles (erythema vesiculosum), there may be a circle of vesicles at the periphery, making the entire spot appear as if ornated by pearls (herpes iris). In rare cases they are discolored by transudation of blood which constitutes the hæmorrhagic form of the affection.

Course.—Gradually the affection heals and fades, leaving a pigmentation behind, after having passed through the usual scale of colors. Normally the affection comes to a favorable termination at the end of from two to six weeks.

Fever does not necessarily occur in this affection, the erythema causing only a slight feeling of discomfort, but morning temperatures of 37.5° C. (98.6° F.) and evening temperatures up to 38.5° C. (101° F.) may occur. At the same time there may be pains in the limbs, and articular swellings. The knee- and ankle-joints particularly are often attacked. Itching or burning occurs only rarely, but more frequently there are pains in the joints. The spots may also spread to the mucous membrane and extend to the lips, cheeks, tonsils, epiglottis and even vulva. Various forms of conjunctivitis have been described by Lipp, von Düring and Fuchs. The involvement of the conjunctiva sets in symmetrically in the form of small spots with a grayish white or yellow surface. They appear slightly raised and extend to the cornea over which they may spread. Renal and intestinal complications (hæmaturia, intestinal hæmorrhages) have likewise been described. Whether endocarditic manifestations have anything to do with this affection, has not yet been proved. In severe cases there may be spasmodic relapses which would protract the course of the disease. Generally, however, severe cases in children are rare, and usually the affection with only slight prodromal manifestations (feeling of lassitude, headache, etc.), reaches its crisis within twenty-four hours, after which gradual recovery sets in.

Anatomy.—The pathologico-anatomical findings consist according to Neisser in a dilatation of the vessels with considerable involvement of the veins, in œdematous swelling of the papillary body and moderate inflammatory migration of leucocytes. The epithelium shows œdematous swellings which are frequently present before the actual formation of vesicles has begun.

Unna, too, looks upon erythema as an affection which has its seat in the papillary body and leads to an œdematous thickening of the same. In consequence of the vascular dilatation there is migration and accumulation of white blood corpuscles around the vessels of the subepithelial network.

Etiology.—Hebra pointed out the frequent occurrence of this disease at certain seasons of the year and indicates particularly October and November, April and May. On the occasion of an epidemic among the army in Constantinople, Düring designated January, March and April as the most favorable times for this disease. Personally I have observed its occurrence most frequently in the spring and fall, although I have never seen epidemics of such magnitude as Herxheimer has described. The frequent occurrence at certain seasons of the year and the clinical course of the disease, rouse the suspicion of its being of an infectious nature, and accordingly the majority of investigators have now expressed themselves in favor of the opinion that erythema exudativum multiforme is of an infectious nature. Neisser believes in miasmatic climatic influences.

The question as to the origin of these efflorescences, whether they are due to a toxic effect from the intestines or whether they are produced by the migration of bacteria, is still undecided. Nor is it demonstrated whether the constipation or intestinal catarrhs which have occasionally been observed in this affection, have any influence on the course.

Differential Diagnosis.—The affection may be mistaken for medicinal exanthema; for the condition of general ill-being described under the name of purpura; for polymorphous erythema; and for chilblains. Mistaking it for chilblains can only occur in the very beginning of the illness, when only a few places on the back of the hands or at the fingers are involved. It is more difficult to exclude toxic erythema, but here also the site, course and concomitant manifestations determine the diagnosis. A large number of the complications attributed to erythema exudativum multiforme (endocarditis, septic manifestations) are probably explained by their having been mistaken for real toxic erythema. Confusion with medicinal exanthema is also possible, but here again the polymorphous character of the latter determines the diagnosis.

Prognosis.—The prognosis of erythema exudativum multiforme is generally favorable. As mentioned above, severe cases are but rarely observed in children. The majority of the malignant cases which have been observed can, I believe, be referred to their having been mistaken for toxic erythema.

Treatment.—The object of the treatment is, in the first place, the removal of the cause, and in the second place the mitigation of the subjective complaints. Thus, rest in bed and lukewarm baths should be prescribed with a view to effecting an improvement in the general condition, thereby relieving the complaints, while internal medication is required to remove any possible intestinal autointoxication. In case of constipation calomel should be prescribed to purge the intestine, or salicylic preparations are indicated to disinfect it. In my hands potassium salicylate, as recommended by Neisser, has given the greatest satisfaction. The dose for children from 1 to 2 years is 1 to 2 Gm. (15–30 gr.), from 2 to 6 years 1.5 to 3.5 Gm. (22–45 gr.), from 6 to 10 years 2 to 4 Gm. (½–1 dr.) a day. Also aspirin and salipyrin (substitutes for salicyl) have met with success. Haushalter and Villemin have seen good results from the administration of potassium iodide.

The external treatment of erythema is confined to washing with spirits (¼ per cent. thymol, 1 per cent. of carbolic acid and menthol, with an addition of 10 per cent. glycerin), and to the application of cooling ointments in order to alleviate the complaints. Cold packs (with acetic alumina) and powdering can also be recommended.

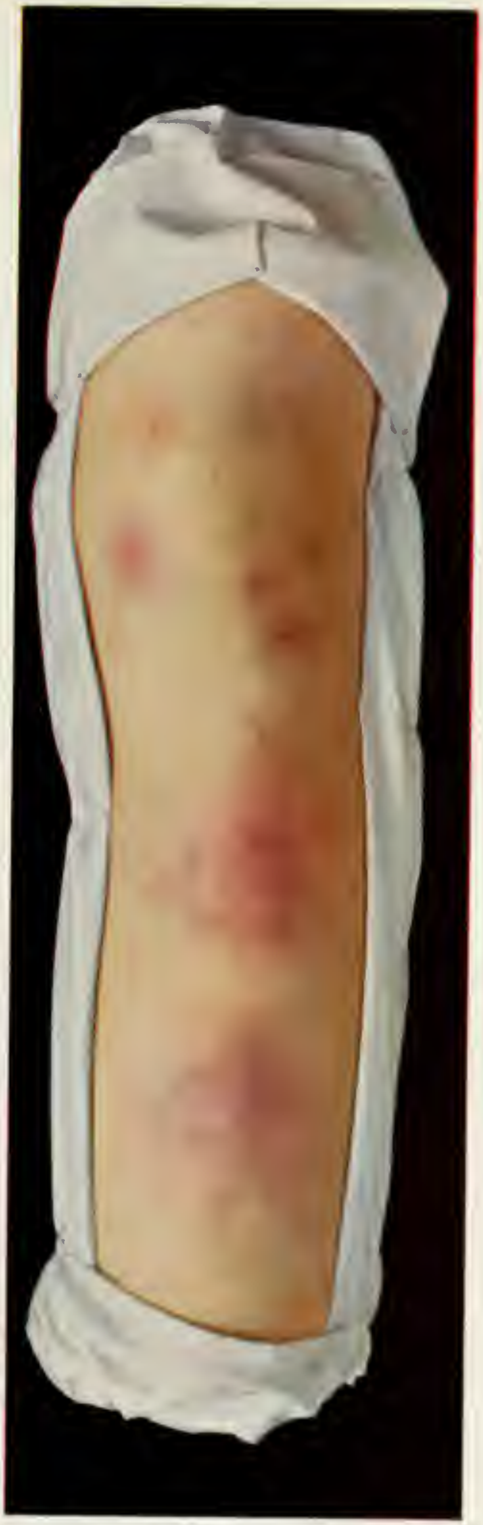
ERYTHEMA NODOSUM

Erythema nodosum is likewise an infectious disease, bears a close relationship to erythema exudativum multiforme and often occurs in conjunction with it, but its course is generally somewhat more severe.

Clinical Picture.—Under prodromal manifestations (discomfort, chills, pains in the limbs, slight elevation of temperature, etc.), nodules of a coarse, doughy consistency are formed in the deeper layers of the skin and in the fat tissue (see Plate 61). They exhibit a bluish red discoloration, are painful on pressure, slightly raised, feel tense, and their size may increase to that of a hen's egg. They first appear on the leg and the dorsal surface of the foot, and may spread to the trunk and forearms.

Course.—The nodules which are subject to change as to their number and size, are gradually resorbed and disappear, leaving a pigment after having passed through the usual scale of colors during the resorption of extravasated blood. Although the number of the nodules is usually between eight and ten, they may increase by spasmodic attacks, in which case the course of the disease would be prolonged.

PLATE 61.



Erythema nodosum.

The nodules generally heal within two or three weeks, but in graver cases several months may pass before the trouble is completely subdued. The fever, which in light cases rises to 38°–39° C. (98.8°–100.4° F.), may reach 41° C. (106° F.), it then gradually recedes—possibly to return again with a fresh attack. Often there are rheumatic pains, there may be vomiting, and the disease seems to present the picture of a severe infectious disease. In these cases hæmorrhagic nephritis, pleuritis, meningitis, endocarditis or pericarditis may develop. Trousseau and Amiaud have observed nodules on the mucous membranes of the lips, palate and fauces. Quite as frequently the affection is said to occur relatively often during convalescence after various infectious diseases, as for instance scarlet fever.

The disease shows a predilection for youthful individuals, but it also occurs in nursing infants, although generally it occurs with greater frequency in more advanced years. Comby mentions 51 cases out of 67 who had passed the third year, while in 16 cases children under three years were attacked. It is equally surprising that the infection more frequently attacks girls. Among Comby's 67 cases 41 were females. In opposition to Kaposi's observation that the affection occurred chiefly in the spring and fall, Comby counted 36 cases from October to March and 31 cases from April to October. There is an interesting communication from Schultheiss calling attention to the frequent occurrence of this affection in Switzerland, his native country, where he had studied it for twelve years, and also to the difference between the temperature curves of erythema nodosum and erythema exudativum multiforme, as against the surprising similarity of the scarlet fever curve and that of erythema nodosum.

Anatomy.—According to Neisser, erythema nodosum is a widely extended inflammation, localized in the connective and fat tissues. The vascular network of the cutis and papillary body is dilated and surrounded by strong infiltration. There is pronounced œdema and great blood extravasation into the tissue. Neisser compares this affection with a hæmorrhagic infarct.

Etiology.—Erythema nodosum is an infectious disease, the virus of which is as yet unknown. Its infecting capacity is comparatively small, as otherwise more small epidemics would surely have become known. Isolated cases of infection from child to child have been described by Para and Moussous, while Abart reported a case of a family epidemic, in which seven children out of nine in one family suffered from erythema nodosum complicated by pneumonia, typhoid, etc. Why erythema should sometimes occur during the convalescent period of scarlet fever, etc., is unknown to us.

Differential Diagnosis.—The diagnosis of erythema nodosum is comparatively easy. It is easily differentiated from boils through the folliculitis which in the latter affection is recognizable in the beginning of the disease, and later through the ulceration. Syphilitic gummata take a chronic course, so does the pathological picture designated by Bazin as erythema induré. It can also easily be distinguished from eminences caused by contusion.

Prognosis.—The prognosis is generally favorable. Here again I cannot help feeling that a large percentage of the reported unfavorable cases belong to general septic conditions complicated by secondary erythema, and not to erythema itself.

Treatment.—The treatment follows that of erythema multiforme. Rest in bed should be prescribed and raising the lower extremities as soon as the nodules appear. In gastric disturbances special diet and laxatives are indicated. The patient gratefully appreciates lukewarm baths once or twice a day. By way of internal treatment salicylic preparations may be given with a view to cleansing the intestine. To this end Boeck recommends antifebrin. The local treatment has to confine itself to reducing the inflammation by cold compresses and to affording protection to the nodules by the application of zinc glue (Leistikow) or ichthyol collodium (Unna). Rheumatoid or articular pains are best treated by salicyl.

TOXIC POLYMORPHOUS ERYTHEMA

Clinical Picture and Course.—Toxic polymorphous erythema is closely related to erythema exudativum multiforme, so much so that the two can often hardly be distinguished. Sometimes it appears in spots, sometimes diffuse, over the entire body or only in parts. It is of short duration and heals comparatively rapidly. It is to be found in the upper sternal region, on the forehead, neck and cheeks.

Etiology.—The causes of this form of erythema may be (1) autointoxications from the intestine, often coupled with extreme constipation; (2) botulism or intoxications from tainted food, etc.; (3) septic processes (ulcerations, etc.); (4) a series of infectious diseases, pneumonia, typhoid, ulcerous catarrhs, diphtheria and severe angina, pharyngitis, affections of the tonsils, etc. In regard to frequency of occurrence, Germain Sée found this form of erythema 12 times in 54 cases of diphtheria, Comby 12 times in 95 cases, Sanné 50 times in 1500 cases. Relatively, therefore, they are not rare. Similarly, Martin de Gimard observed in 1889, 12 cases of erythema in 38 girl patients suffering from typhoid.

Treatment.—The therapy endeavors to remove the toxins and to establish an energetic intestinal desinfection (by laxatives, salicylic preparations, etc.), and in the case of pus foci to interfere surgically. The local and general treatment corresponds with that described for erythema exudativum multiforme.

SEPTIC ERYTHEMA

Cases of septic erythema have been mentioned by various authors (Finger, Haushalter, Singer), in which various bacteria were found in the skin, the erythema and hæmorrhagic foci. This class of erythema has been met with in diphtheria, nephritis, and septicopyæmic conditions, while streptococci in some cases and staphylococci in others were found in the blood as well as in the foci. Neisser and Jarisch look upon this skin affection as part of a pyæmic process. They hold that the skin manifestations are caused by septic bacterial embolisms. For purposes of differential diagnosis it is important to note that this form often occurs combined with purpura, but that on the other hand the stages of development of erythema exudativum are absent.

MEDICINAL EXANTHEMA

Medicinal exanthemata occur in specially disposed individuals in whom medicines produce a toxic effect.

Clinical Picture.—The forms of medicinal exanthema, its appearance and character, are exceedingly variable. They are distinguished above all by considerable polymorphism, so that in one and the same subject we meet with erythematous, urticarial, hæmorrhagic and desquamative conditions and vesicular formations, sometimes interspersed by pigmentations and proliferations of all kinds. It is impossible to establish a typical pathological picture for the reason that one medicine may produce different manifestations in different organisms.

Drug rashes may be caused by external and internal medication. It presupposes a special condition which is designated as idiosyncrasy, that is, a very pronounced susceptibility for a certain substance. Only minimal quantities of such substances are required to exert under certain circumstances a maximum effect. Very often we meet with cases where some chemical substance exercises a cumulative effect upon the body, *i.e.*, that a particular chemical substance is borne very well in the beginning, but that after large quantities have been absorbed, the idiosyncrasy asserts itself. On the other hand it has been observed that acquired idiosyncrasy asserts itself only with certain metabolic disturbances, intestinal disorders, etc., and that only under such circumstances the idiosyncrasy is gradually acquired. Jadassohn has called attention to the phenomenon of immunization: certain parts of the skin which formerly became affected, remain immune on a repeated eruption of medicinal exanthema, so that a partial immunization of single parts of the skin may occur.

In early childhood drug rashes do not occur very frequently in view of the fact that at that period medicines are not frequently prescribed. To facilitate a general survey, I append a brief enumeration of the most frequently used medicines in infancy and their by-effects.

1. Benzoic acid and benzoinate of soda (erythema).
2. Boric acid (erythematous eruptions).
3. Antipyrin, antifebrin, phenacetin, salipyrin (erythema, urticaria, hæmorrhages, wheals, pigmentations).
4. Atropin (scarlatinoid erythema).
5. Arsenic (herpes, urticaria, pigmentations).
6. Tannic acid (urticarial, erythematous eruptions).
7. Bromide preparations (bromide acne, furunculi).
8. Chrysarobin (serious erythematous and eczematous affections).
9. Quinine preparations (erythema).
10. Chloral hydrate (purpura, urticaria).
11. Chloroform (erythema, eczema).
12. Formalin (eczema, nail affections).
13. Iodoform (severe dermatitis of all kinds, erythema).
14. Iodine (nodular eruptions).
15. Opium and morphine, codeine (erythema, urticaria).
16. Phenol (inflammatory dermatitis, erythema).
17. Mercury preparations (erythema eczema).
18. Pilocarpin and syrup Jaborandi (sudamina).
19. Salicylic preparations (erythema, purpura-like affections).
20. Santonin (urticaria).
21. Sulfonal (measle and scarlatina-like erythema).
22. Tar preparations (folliculito-eczematous dermatitis).

Differential diagnosis is not always easy. Sometimes the diagnosis becomes possible only from the frequency of the relapses. Generally speaking, every pathological picture which is remarkable for its polymorphism should arouse suspicion as to a possible medicinal exanthema.

The **prognosis** is benign, as soon as the cause of the drug rash has been established.

Treatment.—The treatment consists in the removal of the cause, in the promotion of diuresis and clearing out the intestine. The local therapy depends upon the stage of the disease, so that it must be anti-urticarial, anti-eczematous or anti-erythematous, as the case may be.

SERUM EXANTHEMA

This form of exanthema may here be only briefly touched upon as it has been dealt with in great detail at another place. The most important form is the exanthema of antidiphtherial serum, which in the first few days has the appearance of urticaria, or between the twelfth and fourteenth days that of roseola or scarlet fever with rather severe general manifestations and articular swellings, fever, etc.

Note.—In tuberculous patients various kinds of exanthema are found upon the skin after tuberculin injection (not to be confused with the reaction in lichen scrofulosorum).

URTICARIAL AFFECTIONS

By the designation of urticaria is meant a pimple, a broad solid elevation of the skin, colored intensely red to white which grows rapidly, may disappear just as rapidly, and itches considerably. The urticarial affections include a group of angioneuroses which occur very frequently in infancy, the most important of which are described in the following paragraphs.

URTICARIA (NETTLE FEVER)

The pathologic picture consists in the appearance of elevated spots or wheals of vivid red color, which often fade off in the centre and itch violently. Their size varies considerably. The wheals may be confluent over a certain area as large as a small plate and even larger, which gives the face an erysipeloid appearance.

The wheals may disappear rapidly, but they may also extend peripherally. We distinguish according to color between urticaria rubra and (if the wheals are white and elevated, of porcelain-like appearance) urticaria porcelanea. Should there, besides, be a formation of vesicles, we speak of urticaria bullosa. Should there be extravasation of blood into these vesicles, we have to deal with urticaria hæmorrhagica.

Course.—The course of urticaria may be acute. There may be a rapid appearance within a few hours and as rapid a disappearance of the entire attack. Very often, however, there may be a recurrence which may take not only a more severe, but even a chronic course. The affection which under ordinary circumstances may be regarded as a light one, now becomes more severe, so that the children are severely taxed by the intense pruritus and the sleeplessness it occasions, and a retardation of growth is the consequence. In these cases the mucous membranes may be involved; the affection may spread to the larynx and pharynx, causing œdema and dyspnœa; even cases of hæmaturia and albuminuria have been described and it is not impossible that even urticarial swelling of the intestine and gastric mucosa may develop. Whenever urticaria becomes chronic, it is a tiresome affection. The relapses recur more or less regularly, and accordingly we speak of a chronic urticaria. Urticaria in infancy is not infrequent and is also observed during the nursing period.

Pathological Anatomy.—The urticarial pimple exhibits a circumscript œdema and a serous saturation of the papillary body and corium with simultaneous dilatation of the lymphatic fissures and lymphatic spaces of the corium.

Pathogenesis.—According to Neisser we have to deal with a vasomotor transudative neurosis, in pursuance of which a more or less diffuse vascular dilatation arises through irritation of the vaso-dilators, in other words an arterial congestive hyperæmia which is followed by swelling, arises from the increased secretion of serum or lymph. Unna believes there is a spasm of the efferent skin veins, and that therefore the development of the wheals is based upon a congestive process. Jarisch looks upon urticaria as the result of two factors acting simultaneously: the general, necessarily central disturbance of innervation of the vessels on the one hand, and the direct lesion on the other.

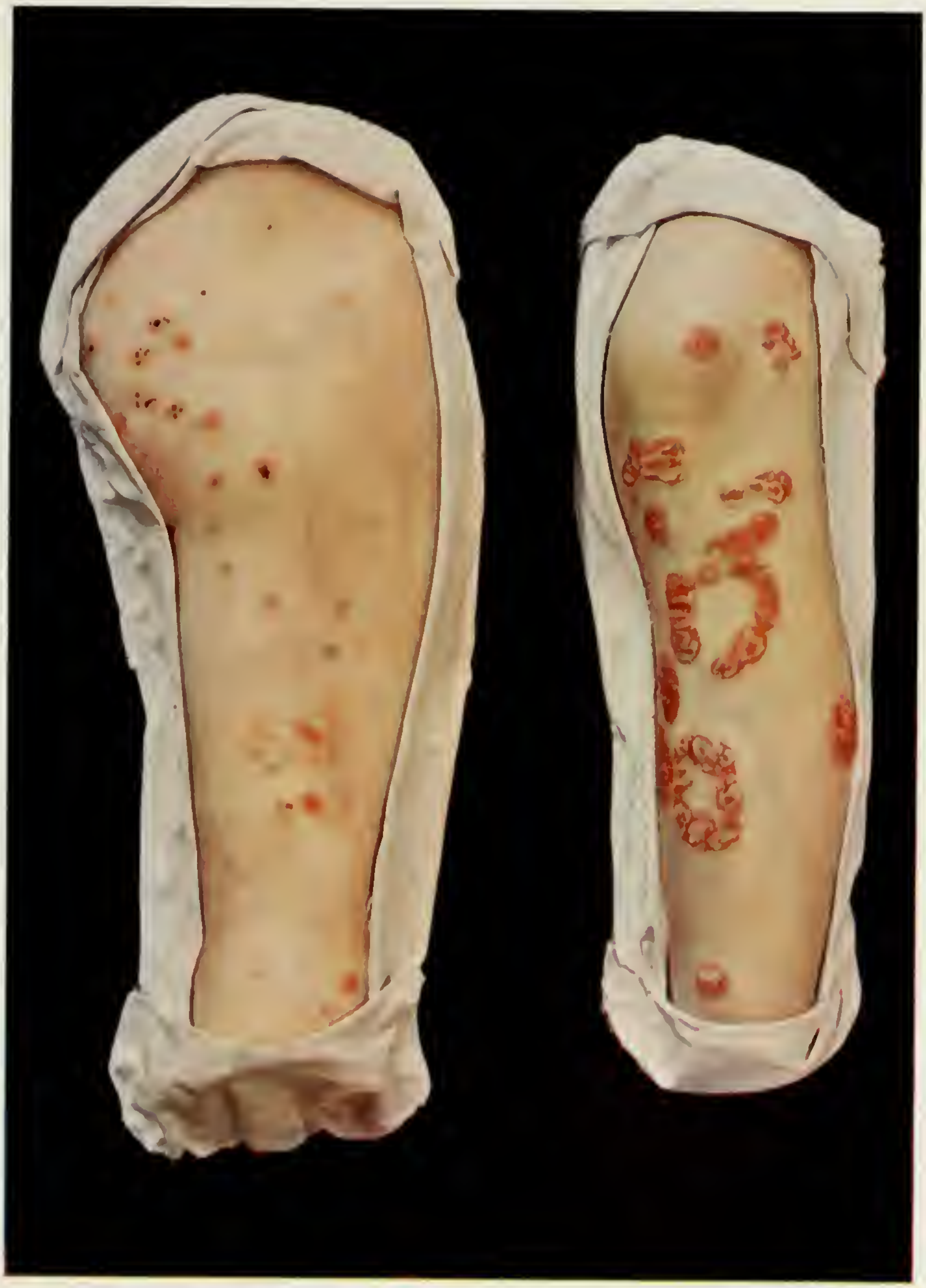
Etiology.—Urticaria may be caused by external irritation (insect bites, sting of nettles, caterpillars). On the other hand it is unquestionably occasioned by substances which affect the skin from the intestine. We thus find urticaria after partaking of strawberries and other fruit, of smoked meat, fish, venison, shellfish, cheese, spices, etc. These substances seem even to be able to exercise an influence on the nursing baby through the milk of the mother, at least Firmin observed urticaria in an infant whose nursing mother had partaken of oysters and fishes. In the third place, autointoxication leads to fermentation and putrefaction (demonstration of indican and sulphuric ether). Parasites in children's intestines cause urticaria, and this seems to be even the case with echinococci. Whether an arthritic process has any influence on the causation of urticaria (through blood dyscrasia) is still doubtful. It is certainly surprising that Wright asserts having cured urticaria by the internal administration of calcium chloride (through increased coagulability of the blood). General nervous disorders like hysteria are a cause of urticaria, especially in young girls. Besides there seems to exist a certain family tendency for the affection.

Prognosis.—In the light form of urticaria the prognosis is very favorable. It suffices to remove the cause, and after a short time the formation of wheals ceases. It is more difficult, however, to effect a permanent cure of the chronic and persistent forms of urticaria.

Differential Diagnosis.—To establish a diagnosis is comparatively simple, as in children only lichen urticarius and erysipelas, or erythema, come in question. The differentiation from lichen will be given in detail in the section which treats of that disease. A confusion with erysipelas and erythema can only occur when urticaria covers large patches of the face or entire parts of the body. The presence of fever will decide the question. In erythema again there is no such extensive swelling and œdema as in urticaria.

Treatment.—The therapy endeavors in the first place to remove the existing harm and its cause. Should the cause be external, protection of the skin should be provided for, while an internal cause should be removed by laxatives and disinfectants so as to cleanse the intestine (calomel, rhubarb preparations, Carlsbad salts in older children; disinfectants: naphthalin, ichthyol and salicylate preparations). In frequently recurring urticaria long-continued use of Carlsbad or Marienbad waters is indicated. By regulation of the diet the intestine should be protected from all irritation. The intestine should be treated direct by irrigation or by washing the stomach.

PLATE 62.



a. Lichen urticatus.

b. Psoriasis gyrata et annularis

The local treatment intends to relieve the itching by washing with cooling fluids in order to prevent the formation of new wheals. Thus, washing with thymol and menthol spirits, acetic water and citric water followed by dusting the skin with talcum or amylum. Besides cooling ointments with menthol or carbolic acid (the doses for little children being quite weak): should be applied. In the majority of cases baths are exceedingly grateful in pruritus, although it happens here and there in children that the wheals increase in consequence of the hot tub. As additions to the bath I have found the following to be the best: soda baths ($\frac{1}{4}$ to $\frac{1}{2}$ lbs. for a child's bath), or sulphur baths with Vlemingkx's solution (30 to 50 Gm. to a child's bath). Also baths of potassium permanganate (3 Gm. per bath) or bolus alba ($\frac{1}{4}$ lb. to the bath) have a soothing effect. If the baths are borne well, children will experience great relief by prolonging them. It goes without saying that nervous or hysterical complaints or disturbances in the general condition should be treated simultaneously, should such be present. To relieve excessive pruritus in older children, it is advisable to try weak doses of antipyrin and potassium bromide.

Urticaria Factitia (dermographisme, dermatoneurose stéréographique, urticaria graphica, etc.).—Gull, in 1859, first described what he called urticaria factitia, being a vasomotor disturbance of the skin recognized long ago. It consists of straight, whealy, white pimples which appear upon the skin on external irritation (scratching, finger nails), they have a red margin and persist for a shorter or longer period. The trouble is mostly met with in persons with a nervous disposition, especially in hysterical patients. It is not very frequent in children. Barthélémy has described it occurring in children of three, five, six and eight years, calling attention to the fact that heredity seems to exert a certain influence.

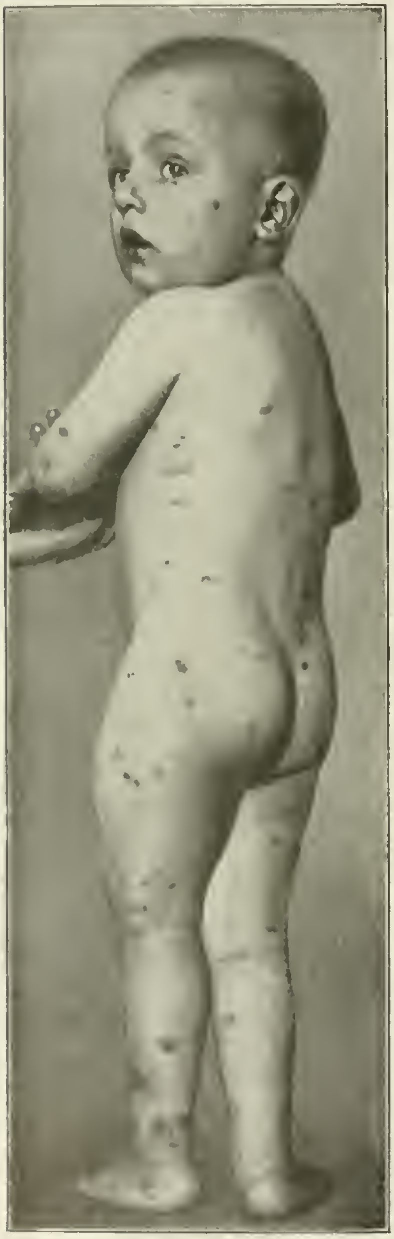
STROPHULUS OR LICHEN URTICATUS

Synonyms.—Lichen urticatus (Villan, Bateman), strophulus (Cazenave), varicellar prurigo (Hutchinson), prurigo simplex (Brocq), prurigo temporanea (Tommasoli), urticaria papulosa (Colcott Fox-Hebra), lichen simplex acutus (Vidal).

Clinical Picture (see Plate 62).—The affection described by Villan, de Rayer, Hardy, and Colcott Fox pertains closely to the urticaria series. It consists of large whealy efflorescences which occur in earliest infancy, disappear more or less rapidly and itch considerably. The wheals become flatter toward the periphery and, in contrast to urticarial wheals, have a conelike, blunt prominence in the centre. The eruption is generally found on the trunk, then the extremities (elbow, knee, flexor surface of the wrist, etc.). Frequently these wheals are covered with a central vesicle, similar to chicken-pox, particularly at the hands and feet.

Course.—The affection generally occurs in the third month, certainly within the first year, and lasts till the third or fourth year, rarely up to the eighth. A few authors (Jarisch) have observed strophulus as early as in the first few weeks of life. The wheals which develop, especially in the evening, are vivid red and itch considerably, so that children show great restlessness prior to an attack and are unable to sleep. After a time the eruption heals until occasionally there is a fresh relapse. The bullous form of strophulus is comparatively not rare (Fox has found 163 cases with vesicles among 209). If the child is taken to the physician in this condition, the vesicles may be able to completely hide the pathological picture, so that it is almost impossible to make a diagnosis. The affection usually occurs during the summer and disappears in winter. In December it is least frequent, in June and July the most frequent. Sometimes it follows in the wake of chicken-pox (hence the name varicellar prurigo — Hutchinson), of measles and dentition (hence the name feu de dentition). Nearly always it is met with in the course of gastric disorders. The papules themselves may last for several days, and as they itch considerably, and as there is consequently a great deal of scratching, they have formed a small scab by the time they come to the healing point. If these spasmodic attacks are of frequent recurrence, the affection may last months and years, taxing the little patients severely by sleeplessness and pruritus. Often the neighboring lymph-glands are found to be enlarged and the skin in consequence of the many scratch effects is in a thickened condition as is also seen in prurigo.

Fig. 67.



Strophulus. (At the lower extremities some of the efflorescences exhibit the bullous form.) Site:—Trunk and extremities.

Pathology.—The affection presents the picture of urticaria complicated by an inflammatory element. This is the central portion of the lesion which grows upon the wheals (Fox-Darier).

Etiology.—The causes of strophulus are almost universally accepted to be fermentation in the intestine, the toxins of which are responsible for the eruption. Blaschko has observed rachitis in 50 per cent. of his cases, and often dyspnœa and dilatation of the stomach. Funk and Grunzach nearly always found rachitis in 45 cases, Blaschko besides found anæmia disproportionately often. Others have held flea bites (Hutchinson) and dentition (Zahnpocken, feu de dents) responsible for it. Personally I have almost invariably found strophulus following in the wake of intestinal disorders and generally obtained improvement and cure by changing the diet. Whether hereditary syphilis can also be held responsible for this affection, is as yet not proven.

Diagnosis.—For purposes of differential diagnosis come in question urticaria, prurigo (see next paragraph) and chicken-pox. From urticaria it is distinguished by the inflammatory cone; from chicken-pox by its long duration, frequent recurrences and nightly exacerbations.

Prognosis.—The prognosis is generally favorable, and the affection nearly always heals spontaneously.

Treatment.—The treatment is in the first place directed to the removal of the cause, and in the second place to give relief to the patient. Therefore, according to prevailing conditions, the first step will be either to attend to the intestinal disorder, treat the rachitis or remove the anæmia. The cleansing of the intestine is done on the principles already explained in urticaria, etc.; in my own cases (and also Zappert's) pure milk diet has proved best. Lassar recommends pulvis liquiritiæ compositus (P.G.) which has also rendered excellent service in a number of my cases. The general condition should be improved by iron preparations and codliver oil with phosphorus.

The local treatment consists in the first place in procuring a cool night's rest and using linen underclothing in order to relieve the itching. Besides, sulphur baths are to be recommended, especially combined with application of sulphur soap foam (Berger's, Unna's, Eichhoff's soaps); also lukewarm salt baths have an excellent effect in some cases. On the other hand, simple warm baths without any addition seem to have an injurious effect. Cooling ointments with carbolic acid (0.5 per cent. caution!) or with 2 per cent. naphthol (Joseph) are applied locally, or cool washings with acetic water and spirits of thymol, menthol or carbolic acid. To relieve the excessive itching Blaschko recommends internally: Antipyrin 1.5 Gm. (22 gr.), aqua destill., syrup, gummos. aa 25.0 Gm. (1 oz.), one teaspoonful in the evening; also baker's yeast (once or twice daily one teaspoonful in milk) or menthol 0.1 Gm. (1½ gr.), ol. amygd. 0.25 Gm. (4 gr.) three times daily 1 capsule) is to be recommended for older children.

URTICARIA PIGMENTOSA (*Sangster*) or URTICARIA XANTHELASMOIDES (*Tilbury Fox*)

Clinical Picture.—Urticaria pigmentosa develops either at birth or during the first year of life, and persists during a great part of the patient's life. It may even develop in utero (Arning, Fabry, Raab) and consists in the appearance of vivid red elevated urticarial foci, roundish in shape. The color of the spots gradually changes into brownish red to sepia brown or yellowish; sometimes brown or sepia brown elevations remain behind, which may be arranged in crests, or in streaks, and represent a tumorlike molluseiform type (Nobl). The affection is found on the trunk, extremities, head and face.

Course.—The eruptions may occur very frequently, especially during the first year, they then become gradually less and disappear toward the twentieth year. One solitary case has been observed to last beyond the fortieth year. Sometimes there is a slight irritability of the vasomotors, frequently erythema and wheals in the old pigmentary places, in other instances the urticarial manifestations are entirely absent and the affected places rather make the impression of a tumorlike elevation up to 1 cm. above the level of the skin. Here and there some of the efflorescences exhibit vesicular formations. Arning and Veiel have observed eczema among the complications. Urticaria factitia has been observed more frequently.

Pathological Anatomy.—Urticaria pigmentosa is caused by the excessive accumulation of those forms of connective tissue cells in the subepithelial tissue layer which have been described by Ehrlich as "Mastzellen" (Unna).

The **etiology** is unknown. Perhaps there is an inherited anomaly (Neisser) or a congenital tendency which proliferates only after birth.

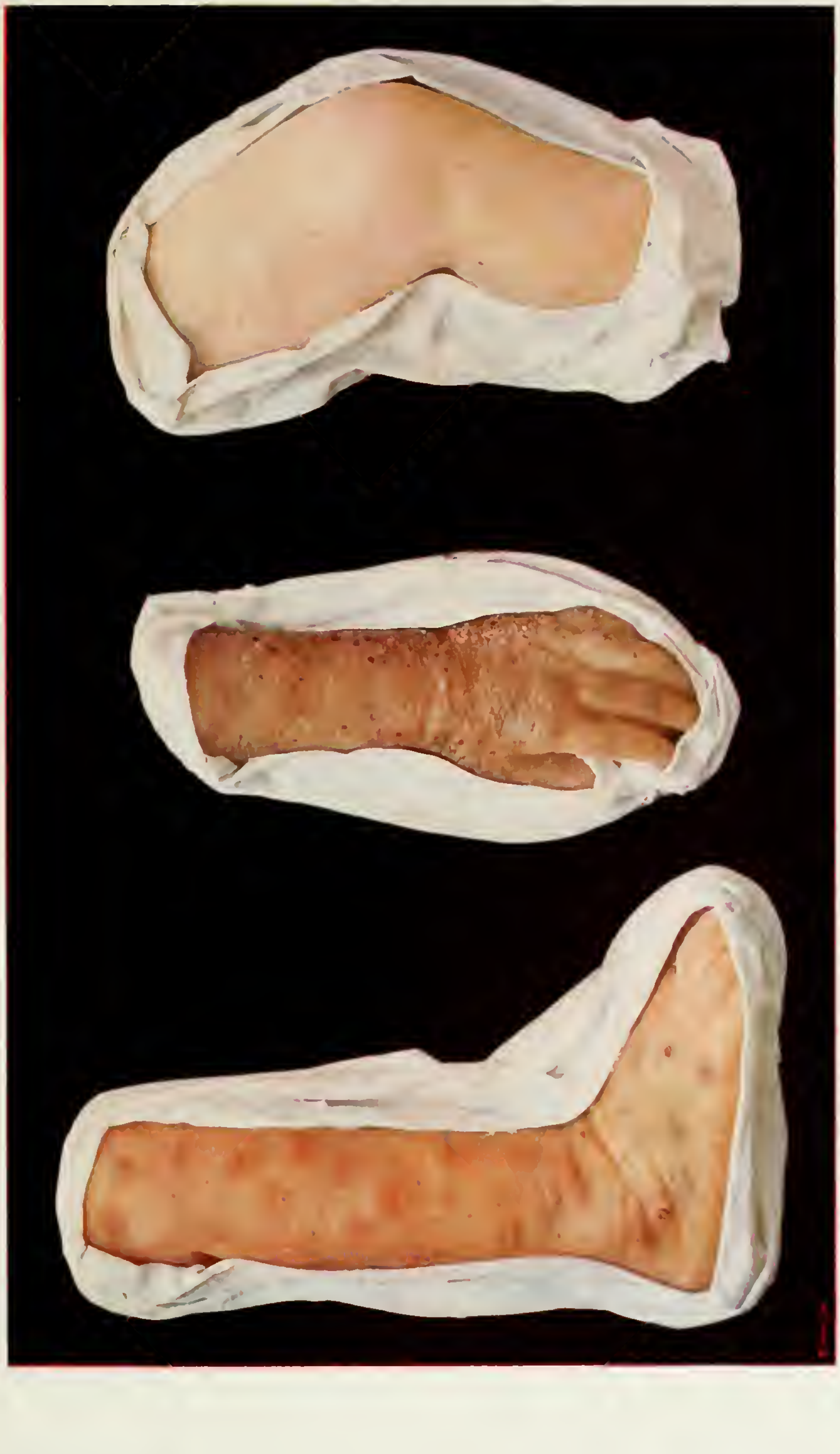
Diagnosis.—For purposes of differential diagnosis there is only urticaria hæmorrhagica (with pigmentary healing) to be considered; indeed there is no doubt that among the one-hundred published cases quite a number belong to that affection. Certainly all cases which have not originated in the first year of life should be regarded with doubt. The affection may also be mistaken temporarily for papulous syphilis.

Treatment.—So far, therapy has proved absolutely powerless. Arning has effected a cure in one case by the addition of 1.5 Gm. (22 gr.) sodium salicylate to the daily ration of milk.

PRURIGO (*Hebra*)

The form of prurigo which Villan has included among the papulæ, has been established as a pathological type by Ferdinand Hebra. It is an affection which begins at the end of the first or second year, attacks the extensor surfaces of the extremities, is accompanied by violent itching and is distinguished by its chronic course. The efflorescences consist of small papules, the size of a pin-head or hemp-seed, which itch

PLATE 63.



a. Prurigo ferox.

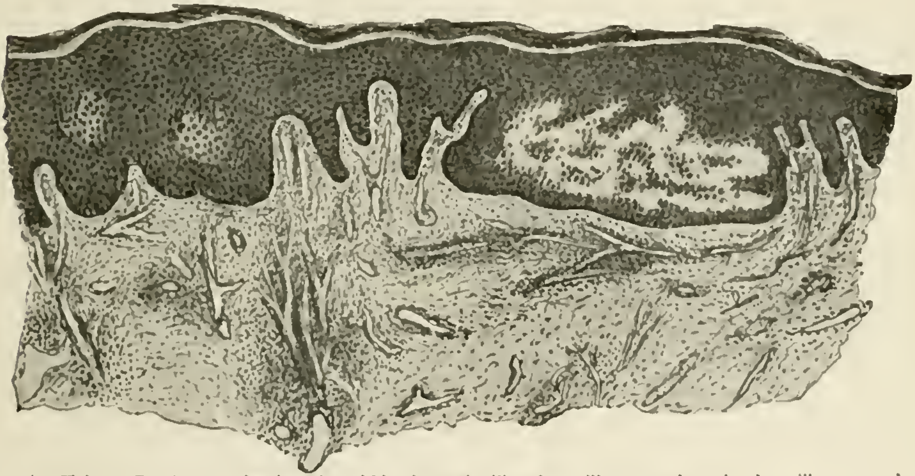
b. Prurigo ferox.

c. Prurigo mitis acuta.

considerably. Their color may be that of the skin or pale red to white, and on account of their tough consistency and localization within the cutis are often more perceptible to the touch than to the eye.

Course.—The affection often commences in earliest infancy with spasmodic urticarial eruptions and lasts longer than ordinary urticaria. It is only toward the end of the second or the beginning of the third year that the small prurigo papule appears, first at the extensor surface of the lower, then of the upper extremities, in the face (rare) and at the trunk. They itch considerably and are therefore usually scratched open and covered by a slight scab. These papules feel like a grater when rubbing the skin, and there is a striking roughness of the skin as contrasted with the smoothness of the flexor surfaces, especially of the articular flexions which are soft and smooth and free from papules. In quite a number of cases, however, these premature urticarial eruptions are absent and the true prurigo papules appear immediately in the first few years. The itching is exceedingly violent. The children are rubbing day and night, and for that reason there are scratch effects in all stages; in prurigo of long standing there may be (as secondary affections) eczema, impetigo, blood excoriations, pigmentations and elephantiastic thickening of the skin. The lymphatic glands, too, especially the inguinal ones, are considerably swollen and known as the so-called prurigo buboes. According to whether the itching is more or less accentuated and according to whether the relapses occur more or less frequently, we distinguish between *prurigo mitis* and *prurigo ferox seu agria.*

Fig. 68.



Prurigo Hebra. Prurigo papules, lymph and blood vessels dilated, papillary vessels and subpapillary vascular network surrounded by small-celled infiltration.

The **course** of prurigo mitis is considerably milder. Gradually the paroxysms cease and under proper attention and treatment the efflorescences heal, the children recover from the pruritic attacks and the trouble is gradually overcome.

In prurigo ferox the relapses follow in rapid succession, the general condition of the child suffers from the attacks, it sleeps badly, becomes nervous and irritable. and generally remains puny, pale, thin, and badly developed. In consequence of the frequent relapses the skin is greatly thickened, has a dark appearance from the many scratch effects and blood extravasations, and on it we find all stages of prurigo, fine small white cicatrices surrounded by dark areolæ, fresh prurigo papules, fresh scratch marks, impetiginous vesicles, eczematous changes. In such cases the entire body and face are usually involved.

The affection is met with in all classes, the lowest and the highest. It usually appears in winter, and although it has been observed by Ehlers and Dubreuilh during the summer, the majority of patients have always been observed in the cold months. The number of girls was twice that of the boys. Heredity also seems to play a rôle.

Pathological Anatomy.—The tissue in the fresh papules is softened, the lymph and blood vessels are dilated, the papillary vessels and the subpapillary vascular network surrounded by small-celled infiltration to the vessels leading downward; at the same time small-celled infiltration in the rete. In later stages cystic formations have been found in the corneal and also in the deeper layers by Kromeyer, Darier, Leloir and Tavernier, Unna, etc. A surprisingly large number of eosinophile cells have been found in these papules.

Etiology.—The etiology of this affection has so far baffled us completely. Its causes are the same as those of strophulus, that is, in the first place digestive disorders, putrid intestines, etc. In favor of this assumption may be cited four cases of prurigo in which Finger demonstrated intestinal putrefaction and products of decomposition in the urine, which were cured without any other treatment than internal medication (dietetic and antifermentative). On the other hand, there are children in which this cause is absent and where we are without a cause. Tommasoli and Besnier speak of a prurigo diathésique, without however being able to adduce a sure proof for their opinion. Again the primary commencement of prurigo with urticaria (observed by Kaposi and later by Riehl) is remarkable and again gives rise to various explanations. Personally I am inclined to look upon the various urticarial affections as separate skin manifestations, caused by different poisons, and poisons of different virulence, from the intestine.

Diagnosis.—For purposes of differential diagnosis strophulus and urticaria come in for consideration; also scabies and the various forms of eczema. If there is a suspicion of scabies, their ducts have to be looked for in the skin; if there is prurigo with excessive eczema, prurigo will always remain behind after the eczema has healed. The facts that the articular flexions in prurigo always remain free and that typical prurigo papulæ are present aid in the diagnosis.

Prognosis.—The prognosis is favorable under appropriate treatment. Just in this affection it is of importance whether the parents are in a position always to attend to their sick child, or whether they leave it at home unattended. The prognosis is generally favorable in prurigo mitis, unfavorable in prurigo ferox. Ehlers has communicated with every prurigo patient who had visited the Commune Hospital in Copenhagen during twenty years and has thereby arrived at the following statistical figures: Cured 23, improved 4, not cured 25, dead 7, not found 112.

Treatment.—The object of the treatment is: (1) to cleanse the intestine as soon as there is a suspicion of putrefaction; (2) to improve the general condition by good nutrition; (3) to relieve the subjective complaints and (4) to establish maceration and to remove all the superficial cutical layers. Internal medication for intestinal putrefaction: benzol naphthol 0.50 – 1 Gm. (7–15 gr.) per day, carbolic acid pills 50 to 60 cg. (10 gr.) per day, and the salicylic preparations previously mentioned, or lactic acid. The general condition should be improved by good nutrition, iodide of iron or codliver oil. To relieve the itching, internal administration of antipyrin 5.0 Gm. (1¼ dr.), syrup simplex 25 Gm. (1 oz.), 1 to 2 teaspoonfuls every evening, or massage as recommended by Murrey and Hatschek. To soften and desquamate the skin, baths take the first place. Sulphur baths are the best (with 50 Gm. powdered sulphur or 30 – 100 Gm. Vlemingkx solution, or 50 Gm. sodium hyposulphite for each bath). The effect of the sulphur baths is increased by washing with sulphur soap (Berger's, Unna's, Eichhoff's).

Joseph recommends baths with lye, once or twice a week. Aside from sulphur baths I have been very successful with diaphoretic measures, prescribing wood tea (decoctions of sudorific woods) and syrup jaborandi (proposed by O. Simon; 1 to 2 tablespoonfuls in the tea), or 20 drops of a 1 per cent. solution of pilocarpin. Baths and diaphoretic measures should be administered alternately; after each bath or diaphoretic procedure the child should be rubbed with a weak epicarin or naphthol ointment (Kaposi, 1–5: 100) or with ung. Wilkensonii, or bandaged with a mild tar-sulphur salve. (When the entire body is rubbed over with naphthol or tar, frequent examination of the urine is necessary.)

HYDROA VACCINIFORME (*Bazin*)

(Summer eruption—Hutchinson)

The skin affection described as hydroa vacciniforme by Bazin in 1861 and again by Hutchinson in 1888 as summer eruption occurs during the first year of life and is comparatively rare, there being only fifty published cases up to the present. The affection usually begins in the first few years and has been described as lasting until the thirteenth or fourteenth year, in one case even until the thirty-first year. In spring, under the influence of the sun, the exposed parts of the body (face, neck and back of hands) are covered after a prolonged stay in the sun with coarse little papules on a reddened base which may be covered by vesicles; these turn into crusts, or the centre may sink in producing a vacciniform fossa, or new vesicles may form at the margin. After ten or fourteen days the crusts fall off, leaving fine white cicatrices behind which look like pox marks. The affection always occurs in spring, recurs in summer and autumn and disappears in winter. It is almost exclusively a children's disease; as stated above, only few cases have been reported at a later age (up to thirty-one years).

Pathological Anatomy.—The skin changes which have been described in detail by Mibelli, Bowen, and Möller, consist in a white serofibrous inflammation of all layers of the skin, there being small vesicles in the rete. Besides, Möller found necrosis of the deeper layers of the epidermis into the corium.

Etiology.—The etiology of this ailment is as yet quite dark. All we know is that it occurs in all climates and that both girls and boys are attacked by it in equal proportions. Ehrmann regards the process as a product of the short-waved and chemically active rays of the sun.

Diagnosis.—The diagnosis gives rise to no confusion.

Prognosis is very favorable.

Treatment.—The treatment purposes protecting the skin from the rays of the sun. For this purpose Unna recommends painting the skin with gelatin, Veiel wearing of orange colored and red veils, Hammer the application of quinine salts in aqueous aceticized solution, Möller believes in hardening the skin by more exposures.

Internal and external treatment have remained without success.

BURNS

According to the nature and duration of the burn we distinguish three degrees.

Burns of the first degree are caused in children by scalding with water at a temperature of 160° to 188° F. or by slightly touching hot objects, etc. The skin becomes red, swells, burns, pains, and exhibits the manifestations of simple heat dermatitis. After a few days the inflammation pales off and there is a gradual transition into the normal state.

Burns of the second degree occur in children by scalding with water between 188° F. and 242° F., steam, fire, etc., acting on the skin only a short time. On the hyperæmic, œdematous skin we observe vesicles which may attain the size of a hen's egg and are filled with a limpid or yellowish serous fluid and after a time become ulcerative. On removing the cystic cover, the inflamed, suppurative rete Malpighii is exposed, and it requires one to three weeks until the new formation of the skin is complete. This takes place in the following way. On the red secretory surface arise small whitish epithelial islands which gradually expand and run into each other. The healed places have first a red and then a brownish pigmentation. Burns of the second degree are exceedingly painful, especially after the cystic cover has been removed and the wound exposed to the air.

Burns of the third degree occur through the long continued effect of boiling fluid, burning clothes, contact with molten metals, etc. They lead to necrosis or eschars of the burnt skin, which looks white, black, incinerated or leatherlike and is insensitive. After a demarcation line has been formed (after about three to five days) the sloughs gradually scale off (one to two weeks), the new formation of the skin again takes place from the epithelial islands as in burns of the second degree. If the defects are very deep, the burns heal with an irregular cicatrix, often with very unpleasant ectopia (trismus) and contractures of the extremities. The more extensive the burns, the more unfavorable the prognosis, the general acceptation being that if one-third of the body has been consumed, all attempts to save the patient's life are doomed to failure. In severe burns on an extensive scale sufferers perish under the following manifestations: whereas during the first day or two the temperature is normal or subnormal, fever gradually develops, and under excruciating pain, conditions of excitation, delirium, numbness, epilepsy, cardiac weakness, vomiting, singultus and anuria occur. After one or two more days death ensues in deep coma.

In children, especially the newborn, the most frequent burns are those of the first degree, caused by hot water, the skin of the newborn infant being so sensitive that it responds with manifestations of burns even to a temperature of 37° C. (98.6° F.).

Prognosis.—In children the prognosis is good only in burns of the first degree; the more extensive burns of the second degree very frequently prove fatal. The prognosis of the third degree is directly unfavorable.

Diagnosis.—For purposes of differential diagnosis pemphigus neonatorum and epidermolysis bullosa come in question. To recognize the former affection, the question of contagiousness is of importance, as to whether similar cases have occurred in the same family or in that of the midwife. In epidermolysis heredity is the point to elucidate, but above all in all cases of burns there are always inflammatory manifestations.

Treatment.—The treatment should be anodyne in the first place by shutting off the air and applying cooling bandages. In the second place antiseptic measures should be instituted in order to prevent the spreading of infectious inflammations. The burns of the first and second degrees are best carefully cleansed, washed and powdered dry with bismuth powder or dermatol. An excellent result is attained in burns of the second degree by Bardeleben's bismuth bandages (open the wheals, cover burned places with bismuth bandage and change bandage every eight days). Limewater bandages with linseed oil (aqua calcis, oleum lini aa 50.0, thymol 0.1). To relieve the pain I have found 10 per cent. bismuth-ichthyol ointment to be of good effect. As disinfecting and cooling lotions after the desquamation, the best effect is produced by acetic alumina, boric acid bandages, etc. In deep extensive destructions Hebra's permanent waterbath, 26° to 30° C. (78.8° F. to 86° F.) will prove indispensable. The patient remains suspended in the uniformly heated water by means of a frame or sheet. If the extremities are extensively burnt, extension bandages have to be applied on account of cicatrization. To meet conditions of excitation, collapse and pain, the corresponding general directions have to be applied.

DERMATITIS HERPETIFORMIS (*Duhring*)

Hydroa pruriginosa (Tilbury Fox), Dermatite polymorphe prurigineuse chronique à poussées successives (Hallopeau), Dermatite polymorphe douloureuse (Brocq).

Under the name of dermatitis herpetiformis, Duhring in Philadelphia has described an affection which occurs comparatively infrequently in children. It is distinguished by its many varieties of form but the herpetic (annular) arrangement of its vesicles is characteristic. In this affection are combined all forms of dermatitis (urticaria, erythema, pustules, papulous manifestations along with eczematous ones), occurring simultaneously or separately. The affection is accompanied by slight fever, considerable itching and burning; the efflorescences may heal, but always recur in the same or a different arrangement. There are continually fresh paroxysms, patients scratch the efflorescences open so that secondary inflammatory manifestations (pustulous or impetiginous, etc.) are found interspersed everywhere. The affection is chronic.

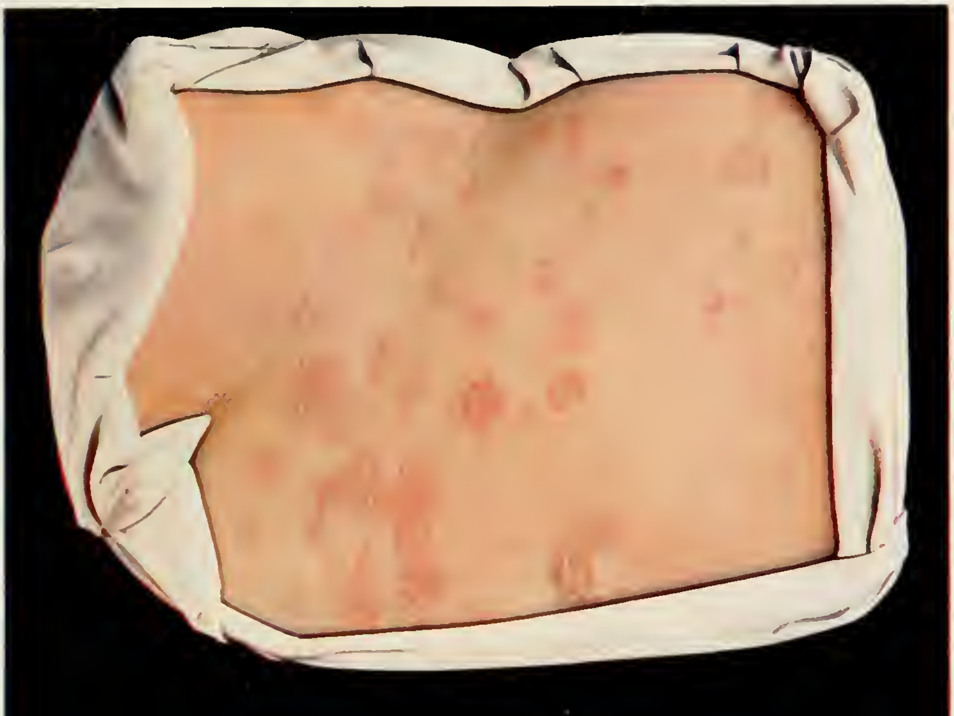
Dermatitis herpetiformis, of which about sixty cases have been reported to have occurred during childhood, occurs chiefly in boys (Meynet et Péhu: 17 boys as against 7 girls), and at every age during childhood. Nearly all patients possess a certain degree of nervousness or are the offspring of nervous parents. Heredity, too, seems to play a certain rôle. Nothing definite is known as to its connection with vaccination.

Etiology.—The etiology is unknown (intoxication neurosis?)

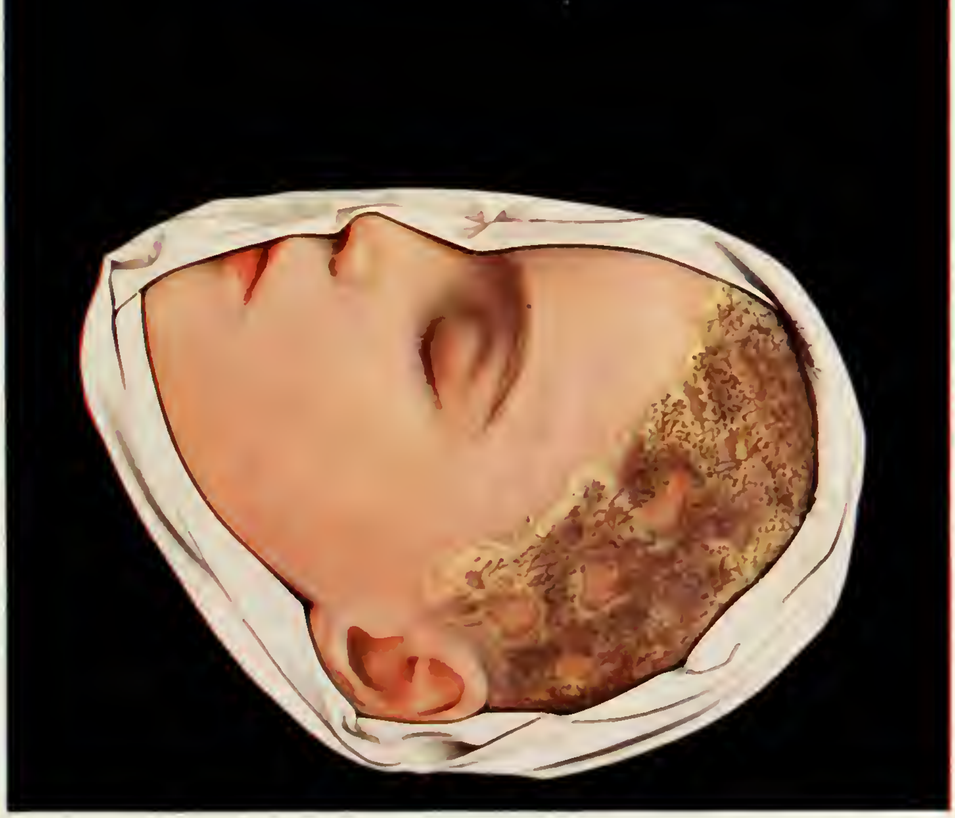
Prognosis benign, aside from chronicity and difficulty in curing the disease.

Treatment.—By the administration of arsenic and strychnine, general dietary rules and due attention to the intestine, the general condition should be improved. Baths and medicaments to relieve the itching should be prescribed (sulphur, tar, etc.); the affected part should be bandaged in order to effect the healing of the dermatitis.

PLATE 64.



a. Seborrheic eczema.



b. Favus.

ECZEMA

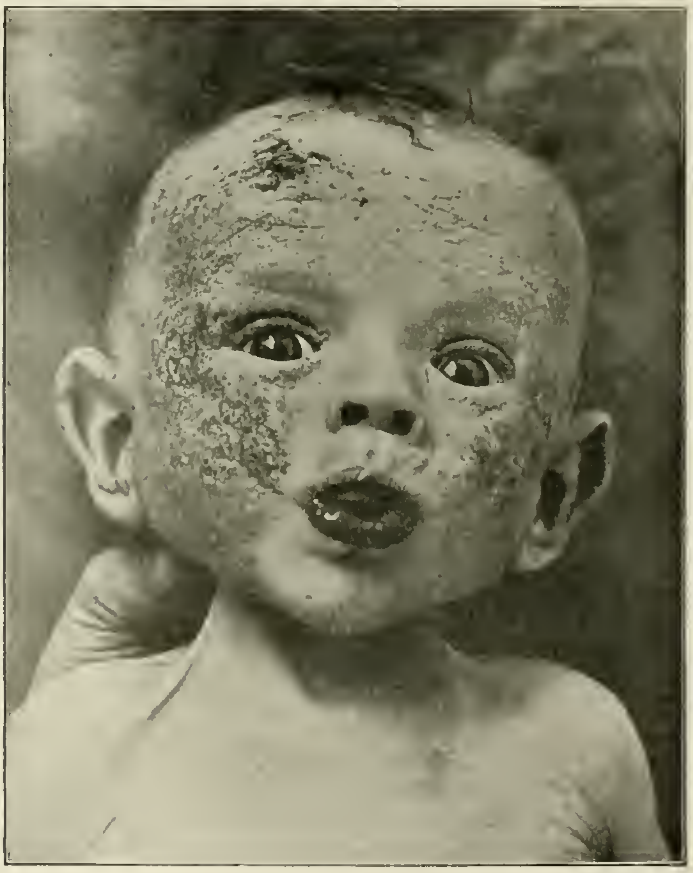
Eczema is essentially a superficial catarrh of the skin, with an inflammation of the upper layers of the connective tissue and epithelium of the epidermis, and is attended by perspiration; there is therefore a surface infiltration and transudation of the corium and vesicle formation of the epithelium. The affection is characterized by acute onset, frequent generalization, extreme irritability of the eczematous skin, tendency to relapses and chronic stage, and healing without leaving any scars (see Plate 65).

The **course** of the affection is generally as follows: At first the skin becomes slightly erythematous and œdematous, tense (*stadium erythematosum*); then small, more or less hyperæmic, not very coarse nodules appear on the skin, which collect around the sweat-glands and hair follicles (*stadium papillosum*). Gradually the small nodules develop into vesicles (*stadium vesiculosum*) and become ulcerative as soon as an infectious stimulus penetrates into the vesicles (*stadium impetiginosum*). If they are opened through scratching and rubbing, and the base of the vesicles is exposed, we observe the hyperæmic, exudative, eczematous layer of the skin (*stadium madidans*). This hyperæmic skin exudes a highly serous, honey-colored fluid, which dries and collects on the skin, forming crusts and scabs (*stadium squamosum*). The longer the affection persists, the more intense becomes the inflammation of the corium and cutis; the skin becomes coarsely infiltrated, highly irritable and responds immediately to irritation by the formation of new vesicles.

Acute eczema occurs in localized, sharply demarcated foci of a papulous and vesicular nature. Soon new foci are being formed in the neighborhood which become confluent and spread: in this way they may involve the entire body, but considerable areas may escape. There is slight elevation of temperature with considerable pruritus which occurs in paroxysms at day and night, causing patients to scratch themselves to such an extent as to expose the exuding skin. After the acute stage has passed into the exudative stage, it may persist for weeks and months, gradually becoming squamous, or the skin may gradually heal so that on superficial examination it appears healthy; nevertheless it is susceptible to every fresh irritation, causing new eruptions and exacerbations at the old spots which had apparently healed. In chronic cases the entire body is often attacked, and the general condition suffers not only from the excessive itching, but also from the sleeplessness it occasions. There arise, especially in children, disorders of nutrition, vomiting and diarrhœa, all of which probably being connected with the eczema; there may be bronchitis and frequently even asthmatic attacks which may occur vicariously with fresh eczematous eruptions. Infantile eczema is divided into attacks during the nursing and later periods.

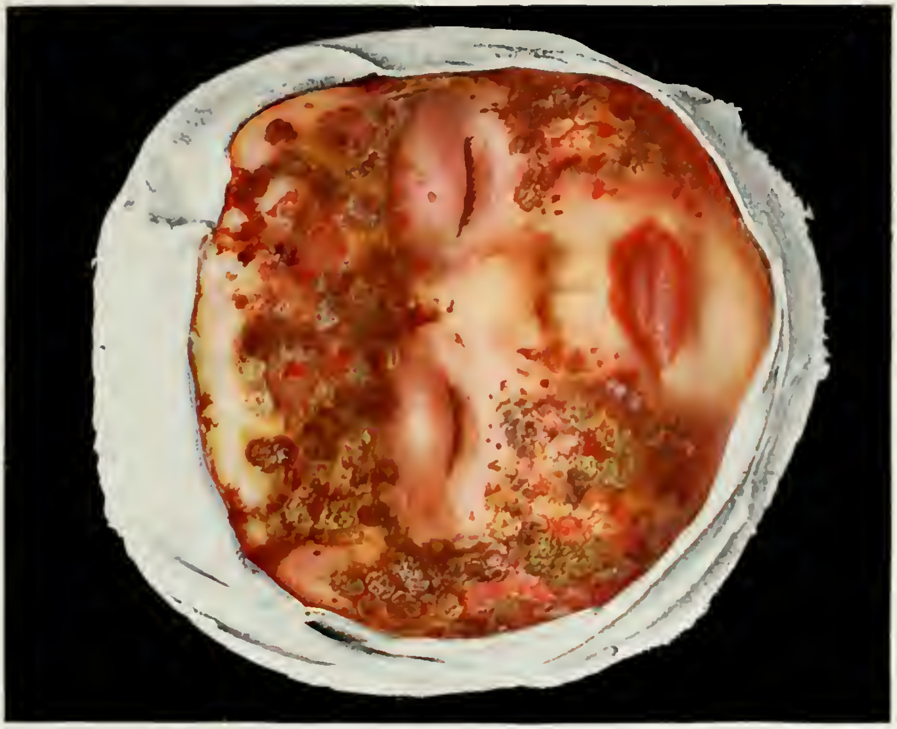
The eczema of the *nursing period* very often begins during the first few weeks of life, and has been observed by some authors (Dubreuilh) as crusta lactea as early as the beginning of the third week. Commencing at the head, face, cheeks, ears, it slowly spreads to the nape of the neck, the chest, and may involve the arms, legs and abdomen. This eczema of the nursing period causes very considerable itching, and children are apt to scratch the pustules open very promptly. At first an erythematous and papulous eruption, it soon becomes vesiculous and remains in a crusty and impetiginous condition for a considerable time because infected from the continuous scratching to which children are prone. In such cases the affection attacks the entire body in the shape of plaques, spreads to the back, chest and extremities, proceeds in the shape of eczematous intertrigo to the genito-crural folds of the perineum and anus, or travels to the cervical folds and articular flexures. This condition remains stationary for many weeks and months; it is a torment not only to the children, but also to parents and physicians. The

Fig. 69.

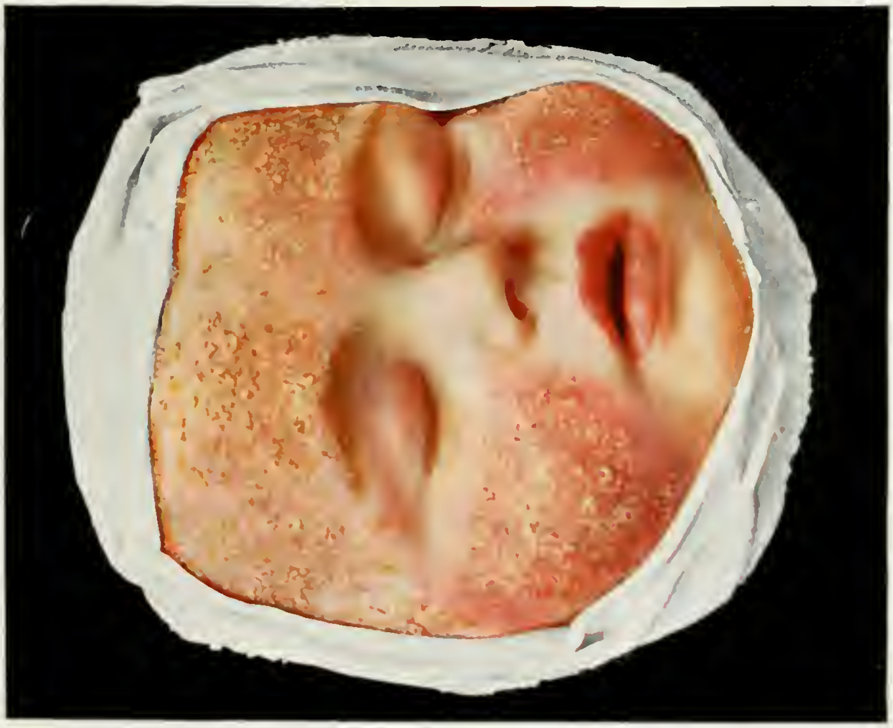


Subacute facial eczema of nursling (stadium crustosum).

PLATE 65.



b. Eczema impetiginosum.

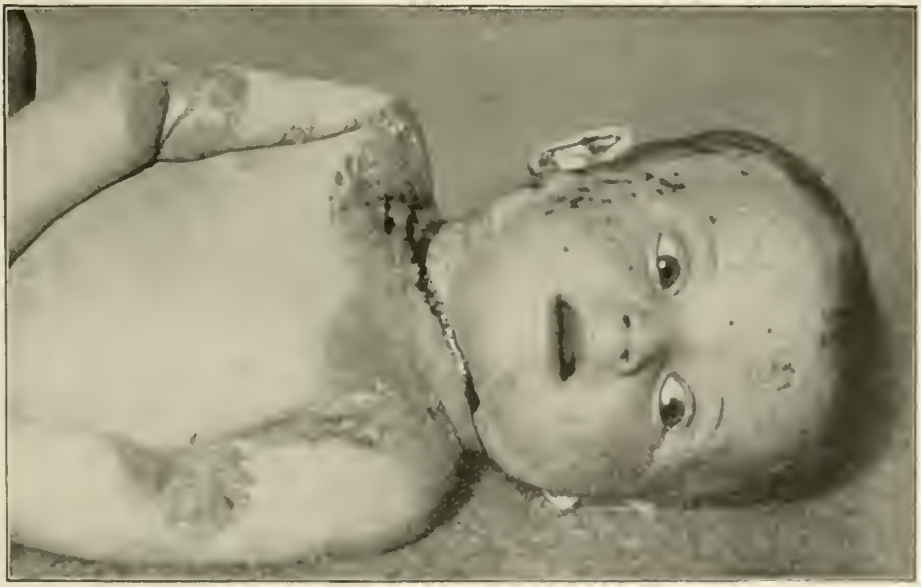


a. Eczema madidum et crustosum.

extent to which nurslings suffer from eczema, varies: especially in the localized trouble children are frequently observed to be of very good general appearance and to thrive well, and only in the highly exudative forms which attack the whole body is there often a considerable disturbance of the constitutional condition.

Eczema of the head of nurslings is of different kinds: (1) the entire face may be attacked, especially in early infancy; (2) sharply demarcated large surfaces of the forehead, cheeks and upper lips may be attacked; (3) the eruption may be confined to small islands of the face; (4) it may originate at the hairy part of the head, when, under the influence of seborrhœa which may often be present, it may spread on. Under the irritation of eczema, together with the honey-colored serous perspiration, with seborrhœic masses, a thick crust is formed, matting the hair together, which emits an offensive odor, and becomes disintegrated. Under this thick conglomeration of hair, crusts, etc., the eczema easily becomes impetiginous, in which case the neighboring lymphatic glands, especially the cervical ones, may be observed to become enlarged and ulcerate. This glandular enlargement as well as the impetiginous changes of the eczema of the head usually spreads symmetrically to the ears, thence to the cheeks and neck, and finally to the entire body. These eczemas of the head and face are exceedingly obstinate and have a tendency to relapse.

Fig. 70.



Eczematous intertrigo. Eczema of the articular flexures.

In a child with scrofulous diathesis we generally find the pathological picture which has been described as *scrofulous eczema.* The nose and the mucous membrane of the nasal ostia are eczematously affected and swollen, and there is œdema of the edges of the lids and lips. The involvement of the upper lip especially frequently causes such an extensive swelling that it assumes a probosciform appearance, and together with the eczema of lids and nose and the scrofulous conjunctivitis which is often present, it often represents the type of scrofulous eczema which may lead to a kind of leontiasis.

The auricles, which become considerably enlarged, are œdematous and crusty, and often, especially in the chronic stage, there are permanent fissures behind the ears at the folds. Very often aural eczema is connected with a purulent catarrh of the middle ear.

Occasionally there is a complication of facial eczema of small children in the shape of a varicellar eczema accompanied by high fever and occurring in sudden rushes, which usually attacks the face first and then spreads to the neck and shoulders. The affection lasts two to three weeks and heals, leaving pigmented spots and flat scars behind which resemble varicella. Kaposi, to whom we are indebted for this communication, has observed one such case with fatal termination. But whether this particular case was not complicated by a secondary varicellar affection, is an open question.

If the eczema involves the trunk of the body, it usually extends first in the shape of *eczematous intertrigo* to the cervical folds, articular flexures, genitals (scrotum or vulva), and then affects nates, thighs and feet. Eczematous intertrigo is caused by friction in the folds and flexures through maceration consequent upon excretions and secretions, and usually occurs in excessively fat children in places where the skin of two parts comes in contact. Intertrigo is accompanied by considerable itching. In this form of eczema we observe all stages of the eczematous process (erythematous, pustulous, impetiginous and similar forms), there are erosions, ulcerations and dermatic changes, which can only with difficulty be distinguished from syphilitic manifestations. For this reason it has been designated by the French erythema syphiloide postérosive or syphilide post-érosive (Jacquet). The forms of eczema occurring at the extremities do not show any peculiarities, but attention should be paid also in these cases to eruptions at the articular flexures.

In the more advanced infantile period two further forms of eczema are of particularly frequent occurrence, aside from the frequently recurring scrofulous eczema. One form occurs especially with the lower class of the population in connection with head lice. As a rule the exceedingly violent itching is responsible for scratches of the skin, a secondary impetiginous eczema developing, which with the crusts and matted hair forms that peculiar tissue which is known under the name of plica polonica, and in which both pediculi and their eggs (nits) are always encountered.

Occasionally an acute eczema with febrile manifestations occurs under the form of *eczema rubrum* which rapidly involves the entire body and is characterized by the phenomenon of the skin of the entire body assuming a bright red to blue red hue, and a scarlatiniform appearance. This is soon followed by the exudative stage, the skin becomes squamous and crusty, and either heals under appropriate treatment or passes into a chronic state, if neglected. The cases are amenable to proper treatment and respond rapidly.

As to so-called *eczematous metastasis,* I cannot bring myself to believe in the occurrence of interior disease after rapid healing of an extensive eczema, although quite a series of observers (Henoch, Comby and especially Gaucher) express themselves in favor of such a possibility. In spite of the relatively large eczematous material which has been at my disposal in the course of years, I have never seen a single case of so-called eczematous metastasis. Nevertheless, it is of course sometimes perfectly possible to observe complications in extensive eczema, such as nephritis; and cases of febrile eczema have likewise been observed (see Prognosis).

Pathologic Anatomy.—This is a catarrh of the superficial layers of the skin; the vessels of the papillary body are enlarged and surrounded by strong small-celled infiltration; the lymphatic fissures are distended; the cutis shows considerable œdematous hyperæmia. Simultaneously there is a swelling of the rete Malpighii and a loosening of the epithelial cells which leads to the formation of vesicles. In chronic eczema there is a still stronger infiltration with new-formations of connective tissue.

Etiology.—Eczema may originate from the most various causes. In the nursing baby we observe in the first place disorders of nutrition. Nurslings' eczema is nearly always encountered in overfed anæmic children with abundant fat cushions, who suffer from gastric and intestinal disturbances, and in whom we have to assume that defective assimilation is the cause of the eczema. The question whether there exists a difference in this respect in children fed on mother's milk and those artifically fed, is not yet determined with certainty. Judging by my own personal experience, I am inclined to believe that artificially fed babies suffer much more frequently from eczema than breast-fed ones. Possibly the composition of the milk of wet-nurses may contain an excess of fat and have an influence on the nutrition of the child. In how far dentition plays a rôle is as yet an undecided question.

Heredity also is unquestionably of some importance, for in some families nearly every child suffers from eczema in its earliest infancy; in the same way scrofulosis and the lymphatic diasthesis—which Czerny treats collectively—are of undoubted importance in the etiology of eczema.

Along with these considerations the state of the skin likewise claims attention. Both excessive dryness of the skin (xerodermia) and an excessively fat condition of the skin (steatodermia) may cause a disposition for eczema. Again, anæmic children who have been badly nursed and fed, are susceptible to eczema.

Attention was called in the chapter on intertrigo to the great importance of the various secretions and excretions in the development of eczema. The secretions of the nose and conjunctiva, the stools, the urine and above all insufficient attention and want of cleanliness are the chief causes of intertrigo. At the same time, exterior causes, application of medicaments, chemical irritation (see medication in exanthema) and thermic irritations (long-continued exposure to sun and heat) will no doubt exert an influence upon the skin. It has also been mentioned that in quite a number of dermatoses (scabies, prurigo, etc.) impetiginous eczema may occur through the spreading of pus cocci.

Since 1891 Unna has taken the position that eczema is of a parasitic nature, describing the affection as an infectious catarrh, and holding the so-called "Morococcus" responsible for the trouble. Likewise Scholz, and after him Raab, have pointed out the frequent presence of staphylococci with the idea of bringing the same into an etiological connection with eczema. I myself, together with a large number of other authors, do not believe in the etiological influence of these cocci, although there is no doubt that for quite a series of infantile variations of eczema we are at a loss for an etiology, and in which we might well think of an infectious cause (see Eczema mycoticum and seborrhoicum).

Prognosis.—The prognosis of infantile eczema is favorable so far as danger to life is concerned. But the question as to whether it is possible in all cases to cure eczema in nurslings and other babies in a reasonably short time, is totally different. Indeed we are compelled to make the sorrowful admission that there are quite a number of infantile eczemas which oppose every external treatment, last for months, and then undergo a spontaneous cure. Generally speaking, however, the great majority of eczemas of nurslings and older children are curable, although frequently an extraordinary amount of trouble and energy has to be expended on the part of both physician and parents. In recent times Feer in a very interesting work points to the frequency of sudden death in eczema and simultaneously to the interesting fact that at autopsy nothing but status lymphaticus was discovered. Feer thinks that the majority of eczema deaths are connected with the status lymphaticus and really belong to the series of fatal cases in that state.

Diagnosis.—The diagnosis of eczema may often be difficult, although in the case of nurslings and older children it is comparatively simpler than in adults. For purposes of differential diagnosis stress should always be laid upon the fact that eczema involves an entire area, that for instance impetiginous eczema extends over a regional surface covered with impetiginous scabs, and that on the other hand impetigo itself is an infection consisting of various isolated foci. The same remarks apply to psoriasis, in which affection the single psoriatic spots can always be traced; they also apply to eczema seborrhoicum, which will be described later on. In favus either the yellow scutula may be discerned or the atrophied cicatrices. It is differentiated from erythema by the inflammation which accompanies eczema; from erysipelas, with which œdematous facial eczema may be easily confounded in the acute stage, by the febrile course. So-called miliaria differs from acute eczema by the superficiality of its appearance.

Treatment.—According to the views which we have laid down in the remarks on etiology, the treatment of eczema is both general and local. The *general treatment* must in the first place take into consideration the etiological factor in eczema of nurslings and of older children. Should a child exhibit the lymphatic or scrofulous diathesis, it will be necessary to try and improve the general condition by appropriate treatment. Similarly, intestinal disorders, constipation, etc., should be energetically treated. In nurslings very great care should be bestowed upon the question of nutrition and regulation of diet. Overfeeding with milk, to which attention has been called by Bohn, Comby, Bellot and Czerny, is also in my opinion without doubt one of the chief causes of nurslings' eczema. I have observed in a number of cases that from the day feeding was reduced, the eczema improved both with unchanged local therapy and with entire cessation of local treatment. It is therefore necessary to pay special attention to the consistency of the human milk, its fat percentage, etc.; in the case of artificially fed babies (who, as we know, furnish the majority of eczema cases) the quantity and consistency (dilution or mixture) should be properly controlled. Also in older children overfeeding has unquestionably an untoward effect, and here again improvement will frequently be achieved by a change in diet, omission of eggs and carbohydrates, giving preference to a mixed diet of milk, vegetables and fruit. The mother's milk, too, should be influenced under given circumstances through appropriate diet. Aside from the question of diet and overfeeding the possibility of underfeeding should not be lost sight of, and attention paid to the connection between eczema and nervous affections (eczema of dentition, etc.). Anæmia should be treated by iodide of iron and codliver oil.

The general treatment by internal medication is comparatively simple. Small quantities of arsenic may be administered in milk in long-continued eczema; in anæmia iron preparations are indicated, and the excessive itching should be relieved by antipyrin (antipyrin 1.50 Gm. (22 gr.) aqua, syrupus aa 25.0 Gm. (1 oz.) 1–2 teaspoonfuls). Besides, in all gastric disorders the well-known laxatives and intestinal disinfectants should be prescribed (calomel 0.01 Gm. ($\frac{1}{6}$ gr.) 3–4 times a day, benzonaphthol 0.2 Gm. (3 gr.) sodium bicarbonicum, magnesia calc. aa 0.25 Gm. ($3\frac{1}{2}$ gr.) nux vomica 0.005 Gm. ($\frac{1}{12}$ gr.) per day according to age).

Local Treatment.—The chief endeavor will be to prevent children from scratching themselves, in which respect they display a marvellous inventive talent. It will therefore frequently be necessary to bandage the hands or to put on gloves after cutting the nails, tie the hands to the bed by gauze strips leaving the arms free to move but not sufficiently to admit of scratching. Similarly under given circumstances cuffs or small splints should be affixed to the elbow-joints so as to prevent flexion of the arms and consequent scratching.

Precaution is needed in washing babies with soap. Generally speaking, moist infantile eczema cases (except perhaps in the very acute stage) can bear baths quite well, but it is frequently found that each washing of acute and subacute eczema cases is followed by irritation and exacerbation. For these cases washing with some spirits ($\frac{1}{4}$ per cent. spirits of thymol and 10 per cent. glycerin) or cleansing the skin with cold cream and vaselin is to be recommended. Baths, generally speaking, may therefore be given, the cleansing of the body alone requiring regular baths in the case of nurslings; especially bran, chamomile or wild thyme baths are borne excellently. The only requisite is that immediately after each bath corresponding medication is applied to the skin (powders, ointments, pastes) and it should be remembered that with children it is not enough to simply put the ointment on the skin but that a bandage should be applied over it. Nearly every child, especially nurslings, would soon rub the ointment off and scratch open the eczema, unless every possibility of doing so has been destroyed by a firm bandage. Eczema will not heal unless the air is shut off and patients are absolutely prevented from scratching the wound open every day, and unless proper medication for each condition has been correctly applied and used for a sufficiently long time. The medication should of course be always adapted to the various stages of the disease.

Thus, in the *erythematous and papillous stage* washing with $\frac{1}{4}$ per cent. spirits of thymol, $\frac{1}{2}$ per cent. spirits of carbolic acid, 2 per cent. spirits of boric acid would be indicated, followed by dusting with powder, which may also be applied without previous washing. The best is mineral powder (talcum with oxide of zinc, vasenol powder), because vegetable preparations easily form a paste with the eczematous exudate. Dry paintings are also to be recommended with either Boeck's or Neisser's preparation (see p. 437) and these are best followed up with powder.

In the *stadium madidans* the treatment will consist in producing an absorbing and antiphlogistic effect by means of moist bandages (acetic alumina 1:10, 3 per cent. boric acid, $\frac{1}{4}$ per cent. resorcin). As soon as the exudation is stopped, absorbing pastes (starch, vaselin aa 20.0 being the cheapest; oxide of zinc, starch, lanolin, vaselin aa 10.0 being the best) should be applied to effect a complete desiccation and improvement of the inflammation. An addition to this paste of 1–5 per cent. lenigallol has an excellent effect in these exudative eczemas. The effect of lenigallol is not only curative, including the inflamed and changed tissue, but also caustic by drying the acute inflammatory places and favoring scab formation of the eczematous surface. In order to minimize the irritation from itching 1–5 per cent. tumenol may be added to the zinc paste, and as soon as the eczema approaches the final healing stage, a very weak concentration of tar may be added (see p. 439). It is a mistake to apply fatty ointments to exuding places, because the serous exudate collects under the fat layer and serves to accentuate the irritation of the skin. In the *crusty stage* the crust should first be softened by an oil bandage and removed, after which the exudative and still hyperæmic and exposed skin should be immediately treated with paste. The *stadium squamosum,* in which the acute manifestations, the exudations and irritability of the skin are becoming less, requires a softening of the thick, chronically infiltrated places by ointments and plasters. Hebra's ointment (or better still unguentum vaselini plumb., which is prepared with equal parts of vaselin and lead plaster, to which 1 per cent. of carbolic acid is added for better keeping): bismuth and zinc aa 1.0, ung. leniens, ung. simplex aa 10.0, Neisser's ointment, and Rille's ointment; salicylic acid 0.5, lanolin 60.0, vaselin 30.0 are all useful. Among plasters I have found Pick-Arning's 2½ to 10 per cent. salicylic soap plaster the best. It will often be necessary in this stage to produce an inflammatory and macerating effect in order to soften old places. For this purpose a 10 per cent. concentration of the above-mentioned soap plaster, also pyrogallus ointments (1–5 per cent., weak chrysarobin ointments) and especially ointments containing tar.

I would still like to discuss the various localizations of eczema in nurslings and older children, together with the corresponding therapy.

In *head and face eczema* of little children an oil bandage should be first applied to the head (the best being of gutta percha or parchment paper, Billroth's batiste) in order to remove the scab formation. For this purpose the head is washed daily with lukewarm chamomile water, well rubbed with (Unna's) tar soap, then with pure codliver oil, and firmly bandaged. Better than simple codliver oil is a 2–10 per cent. mixture of salicylic oil (Neisser), to which castor oil is added to dissolve the salicylic acid (salicylic acid 2-10.0, ol. ricini 10.0, ol. oliv. ad 100.0). This procedure is continued daily, until after the lapse of a few days the scabs come off and the eczematous base is freely exposed. The head is then bandaged for another few days with the oil cap, and later with suitable ointments (Rille's salicylic vaselin or Neisser's ointment of bismuth and zinc. In very obstinate cases a weak concentration of tar may be carefully added to the final applications of these ointments. But for a long time afterward eczema of the head requires energetic care and attention by keeping the scalp in a fatty condition.

Exudative *facial eczema*, especially of nurslings, requires most diligent application and careful bandaging. Here again the exudate is to be removed first by moist bandages (especially at the œdematous swellings) and then dried up by zinc pastes (to which lenigallol or tumenol may be added according to requirements). The desired effect having been accomplished, the pastes may be gradually changed to ointment by the addition of olive oil, so as to soften the skin. An excellent effect is here produced by Lassar's zinc oil, oxide of zinc, ol. oliv. aa 50.0 (or Schlossmann's paste of oxide of zinc and vasenol) in the place of the paste and oil. Should, however, the facial eczema continue to exude, it is advisable to stop the exudation by slight caustic applications either of lenigallol paste or of nitrate of silver. The painting with silver (Burchardt) has an excellent effect upon the exudation through the formation of silver chloride. The exuding fissures are blocked up, the exudation ceases, and if this procedure is repeated daily for 3 or 4 days, and later every other day, the usual result is a dry skin in the second week; the treatment may then be continued with paste. While the painting is carried on, it is advisable to apply powder or paste. As soon as the skin is dry and there is reason to believe that there will be no more exudations, ointment will also in these cases take the place of paste. In these conditions Ehrmann recommends a 5 per cent. xeroform ointment; personally I have seen the best success with Hebra's ointment, Neisser's ointment of zinc bismuth leniens and Wilson's zinc oxide ointment. Where there are old places to be softened, careful application of tar will also have to be made. In place of the tar, anthrarobin (10 per cent. solution in tinct. benzoes), proposed by Behrend, or Arning's painting with tumenol 8.0, anthrarobin 2.0, ether 20.0, tinct. benzoes 30.0, can be recommended.

Eczema of the *eyelids* is best treated with yellow oxide of mercury. Eczema of the nasal apertures and folds is first treated with zinc paste and later with ointment, having special regard to catarrh of the nasal mucous membrane, which is frequently responsible to a certain extent for this condition. Here again a bandage should be applied, so far as may be possible. Frequently eczema about the mouth is caused by the use of mouth washes; this should be met by stopping any and every kind of mouth washes, after which the usual eczema treatment should be applied.

Particular attention should be paid to eczematous intertrigo. Here again cleanliness by bathing is of importance. During the first stages powder and lint (the best being boric lint) should be applied to the anal and genito-crural folds so as to prevent friction and favor the absorption of the secretion. In exudative eczema, it may also here become necessary to resort to treatment with moist bandages and paste. The open gangrenous and ecthyma-like places which are often present in intertrigo around the anus, are best treated by baths and irrigations. Besides, aristol and dermatol pastes (5 per cent. of which is added to the zinc paste) should be used to promote skin formation and granulation. Also powdering with aristol and dermatol is indicated.

Eczema of the extremities is treated upon the same principles. Very frequently it is advisable to treat a crusty and no longer exuding eczema with gelatin and zinc (see p. 436), because underneath this bandage the eczema heals, while the bandages need not be renewed very frequently.

At a more advanced age eczema occurring in conjunction with pediculosis requires special treatment. The removal of the vermin is best effected by petroleum or 5 per cent. naphthol, in order to ensure a minimal irritation of the underlying eczema. The eczema will then heal under applications of salicylic oil and mild ointments. Cutting off the hair is only necessary where the hair is so matted that it refuses to separate. Generally speaking, the hair should be saved, whenever possible.

Eczema rubrum which attacks large areas of the entire body is best treated by Lassar's cream zinc oil, and later by baths and ointments. In children a hand and finger eczema occurs very frequently between the hands, accompanied by hyperidrosis; this should be treated with hot hand baths of boric acid and weak resorcin paste (½ to 1 per cent. added to the zinc paste).

The question in how far older and chronic eczema, also that in infantile periods, can be cured by the application of Röntgen rays, has not yet been sufficiently tested.

ECZEMA SEBORRHOICUM (*Unna*)

In 1887, Unna described an affection of the skin under the name of Eczema seborrhoicum, which was called by Neisser Mycosis seborrhoica, and is frequently mentioned in the literature as mycotic or psoriasiform eczema. This is a pathological condition of the skin which does not belong to the eczematous series proper, and is probably caused by a microörganism. At the same time it may often exhibit eczematous changes, such as hyperæmia, inflammation, exudation, etc. In infancy the affection usually starts from seborrhœa sicca (Gneis) of the hairy part of the head; it is very frequent in little children and will often persist until a more advanced age. Forming perfectly round rings, this eczema spreads from the head to the feet. The various spots are sharply demarcated, dry, slightly itching, with lardaceous appearance or covered with scales and scabs, of yellowish to yellowish red color. The affection may become worse, as the fatty secretion increases; or again the spots may occasionally have the appearance of psoriasis. The determatosis is not infrequently met with in infancy and is usually connected with the status seborrhoicus which will be described later on when treating of seborrhœa; its favorite places are the hairy parts of the head, the forehead, face, sternal region, intracapsular space and axilla.

Etiology.—The parasitic nature of eczema seborrhoicum is very probable, but not yet proved. It requires further investigations to determine whether Unna's morococci or the bottle-shaped bacilli which are frequently found on the scalp, are the cause. There is no doubt that the development of parasites is favored by the maceration and the abnormal secretion of fat.

Prognosis.—The prognosis is generally favorable, but seborrhœic eczema has a tendency to frequent relapses.

Treatment.—If the affection shows dry, easily desquamating areas, the treatment of eczema seborrhoicum consists in the application of sulphur-resorcin ointments (having an antiparasitic effect, also softening and removing the horny masses), often in conjunction with the sulphur—ichthyol and thigenol. A very energetic effect is produced by painting with weak anthrarobin and chrysarobin. At the same time soap treatment. The exudative, irritated places of eczema seborrhoicum are first dried by pastes.

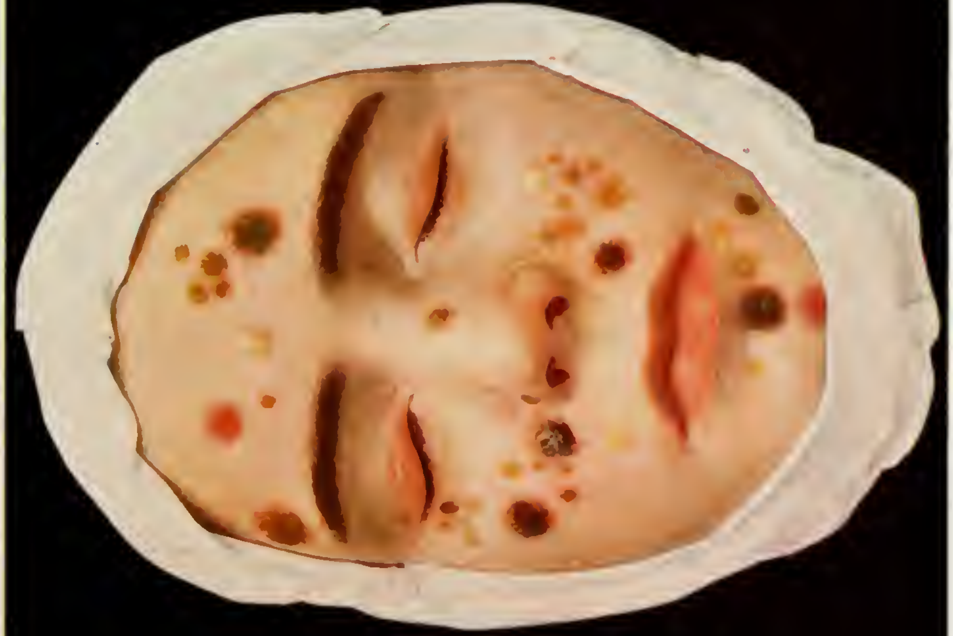
IMPETIGINOUS AFFECTIONS

Impetigo is an affection caused by the inoculation of staphylococci and streptococci, characterized by the appearance of pus pustules. Any eczema, purigo, etc., may become secondarily infected and therefore impetiginous as a result of the entrance of these cocci. The following forms may occur in the periods of infancy and childhood.

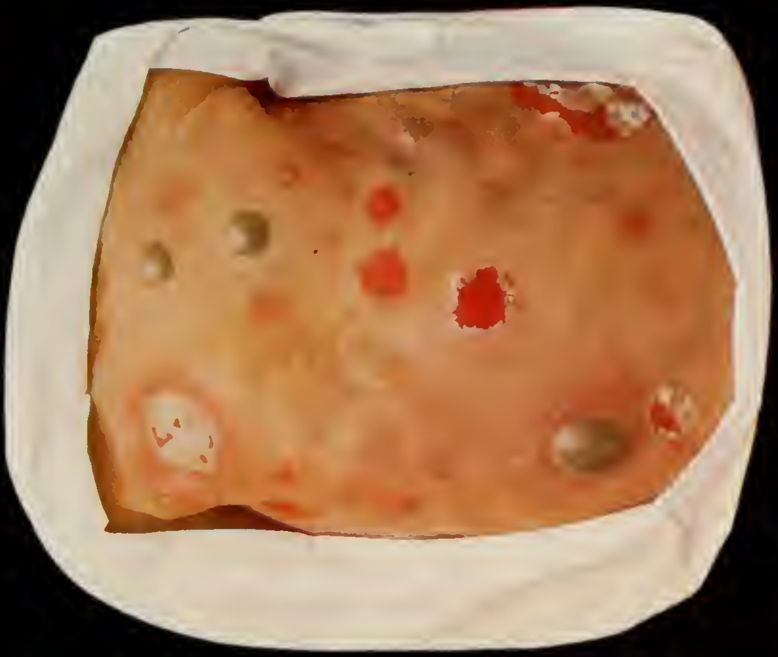
IMPETIGO CONTAGIOSA (*Tilbury Fox*)

This affection was described in detail by Tilbury Fox in 1864, and consists of small watery vesicles underneath the corneal layer which may gradually attain to the size of a dime to a silver dollar (see Plate 66). They are flabby, filled with seropurulent fluid, and surrounded by a slightly inflamed areola. The contents of the vesicles soon become purulent, the vesicle bursts and the spot is covered with a yellow scab which looks as of it were glued on (Fox). The vesicles stand separately, often very near each other, and may coalesce. After removing the crust which was formed through the bursting of the vesicles, a pale red moist surface which may rapidly heal or extend peripherally according to circumstances, will be disclosed underneath a thin purulent coat.

PLATE 66.



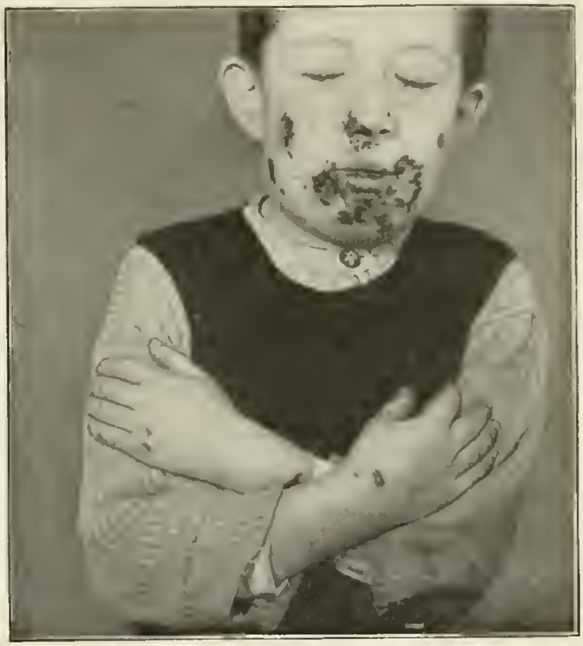
a. Impetigo contagiosa.



b. Pemphigus neonatorum.

The affection is met with in children of all ages, who are principally affected in the face, on the hands and the hairy part of the head or at any other part open to contact; through scratching it is transmitted also to the covered parts of the body. By healing in the centre and slowly spreading at the margin, it may assume circinal and annular forms (impetigo circinata). On the mucous membrane, too, aphthous forms have been observed by Jadassohn.

Fig. 71.

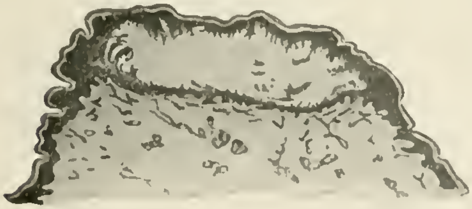


Impetigo contagiosa. Typical site around the mouth, circinal form at the left angle of the mouth

Impetigo is a contagious disease. Quite a series of epidemics have occurred in schools, institutions, etc., and it has also been observed as a sequel to vaccination (for instance in 1885 on the island of Rugen).

Pathology.—The corneal layer remaining intact, there is formed a vacuola in which a fluid accumulates containing more or less pus corpuscles, and which proves to be a serous exudate. The rete underneath the vesicle is widened, the cells of the upper stratum are swollen, those of the lower stratum are normal. The papillary vessels are dilated, also those of the cutis, and there is a slight superficial infiltrate.

Fig. 72.



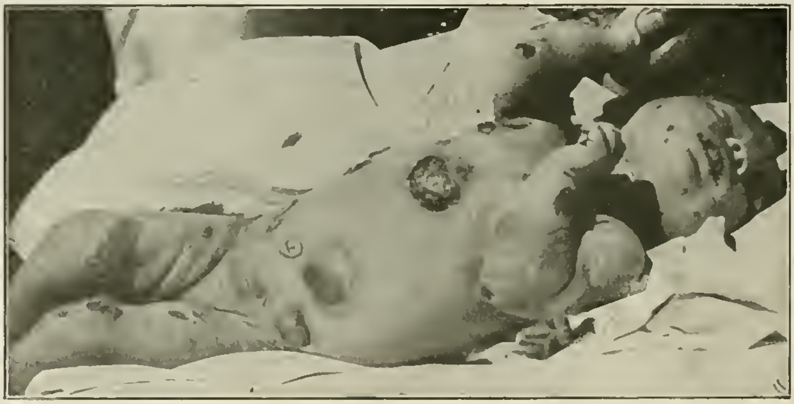
Impetigo pustule. Corneal layer intact, vacuola with leucoserous exudate (in this specimen dropped to a large extent) papillary vessels dilated, slight infiltration.

Etiology.—The affection, which is exceedingly contagious, is probably caused by streptococci (the most infectious kind) and the less infectious staphylococci. In numerous cases these microörganisms have been transmitted into the injured integument. It also appears that there is a connection between impetigo contagiosa and pemphigus neonatorum.

Diagnosis and Prognosis.—The differential diagnosis is not difficult, as in the case of children only chicken-pox (without any red inflammatory areola) and impetiginous eczema (involvement of an entire area) come in question, and the isolated efflorescences in impetigo are always present to secure the diagnosis. The prognosis is favorable.

Treatment.—The treatment consists in opening the vesicles, removing the crusts and cleansing the skin. To this end we avail ourselves in the first place of the sulphur preparations which have an excellent (almost specific) effect on all impetiginous affections. Therefore, as the case may be, the vesicles should be opened first or the crusts softened with oil, after which the affected place should be bandaged with sulphur zinc paste or sulphurated boric vaselin (10 per cent.). At the same time washing with sulphur soap is indicated, or sulphur baths (30 to 50 Gm. Vlemingkx' solution) if the trunk should be involved. Jarisch favors a 2 per cent. mercurial salve (ung. hydrargyri cinereum P. G.). The body linen should be regularly changed.

Fig. 73.



Pemphigus neonatorum. Cystic remnants after bursting of the vesicles.

PEMPHIGUS NEONATORUM

Synonymous.—Schalblasen, Schalblattern, pemphigus infantum (Escherich, Rille), pemphigus neonatorum epidemicus (Unna), pemphigus contagiosus (Faber), exanthemata bullosa neonatorum (Bärensprung).

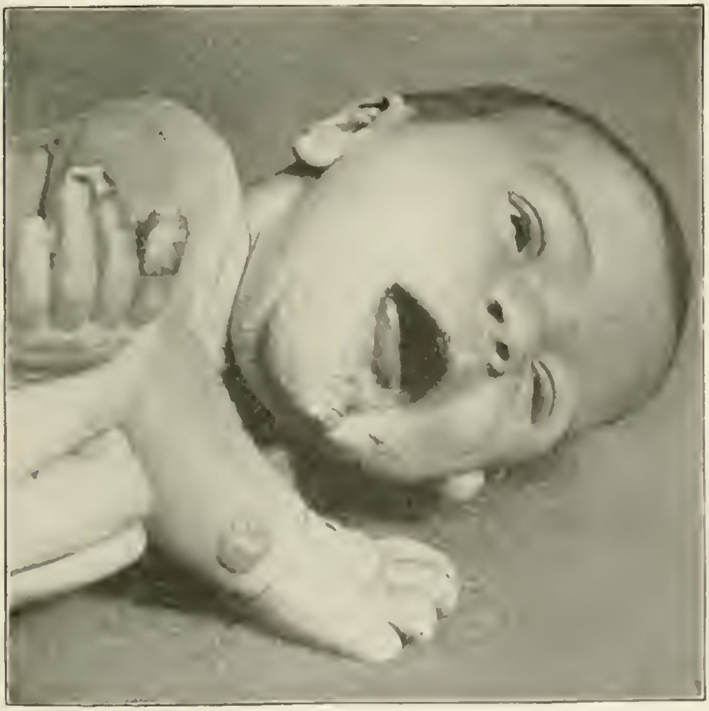
This affection was first described in 1610 by Forestus as dyscrasia and up to 1870 or longer was looked upon as cachexia; only in the last thirty years its bacteriological cause was recognized.

Clinical Picture.—Between the third and eighth day there is a sudden appearance of coarse cysts from the size of a pea to that of a hazel-nut, which are at first tense but finally become flabby and burst (Plate 66). The fluid they contain is at first limpid, then clouded. The skin is apparently normal or at most slightly hyperæmic. As the cyst bursts, a red exudating surface remains which is covered by the margin of the epidermis and is promptly renewed. The affected spots remain hyperæmic for some time. The cysts appear at all parts of the body (rarely on palms of the hand and soles of the feet); they may keep recurring for a time and thereby assume a more chronic form which occasionally renders the affection much more malignant and fatal. As a rule the affection which is usually afebrile, is a light one and cured in five weeks or less.

Pathology.—According to Luithlen the cyst formation occurs by an elevation of the corneal layer; the vessels of the corium are dilated and surrounded by numerous leucocytes. Staphylococci are principally met with in the contents of the cyst.

Etiology.—The affection is contagious and quite a series of epidemics and transmissions to children, mothers and nurses (family epidemics) have been reported. If vaccination has taken place, hyperaemia occurs five to seven hours later, after twenty-four to seventy-two hours the pemphigus cysts develop, in which according to the bacteriological investigations of Almquist, Strelitz, Escherich, Peter and others, the principal bacterium is the micrococcus aureus, but others are also met with. But since Almquist and Strelitz were able to produce pemphigus cysts by inoculation with the staphylococcus aureus, we have in all probability to regard this coccus as the causative factor. It also appears that there is a connection between impetigo contagiosa and pemphigus neonatorum. According to investigations made by Nobe, Richter and Matzenauer, impetigo contagiosa can be produced by inocu-

Fig. 74.



Pemphigus neonatorum. Flabby cyst.

lating the contents of the cyst upon older children. In the reverse way, impetigo contagiosa can be transmitted to newborn infants producing pemphigus neonatorum; or at any rate, in both affections the same staphylococcus is met with and dermatitis exfoliativa seems to have the same etiology.

Diagnosis.—There is no difficulty in establishing the diagnosis of pemphigus neonatorum.

The **prognosis** is generally favorable, although several epidemics have been reported in which there was abnormal mortality (Huart 1878, 60 per cent.; Corrigan 1834, 90 per cent.). In these cases, however, the victims were invariably badly nourished foundlings. In all cases of pemphigus with unfavorable or fatal termination there is always the possibility of pyæmic affections in which the skin shows only part of the disease, or cachexia may be the cause of the fatal termination.

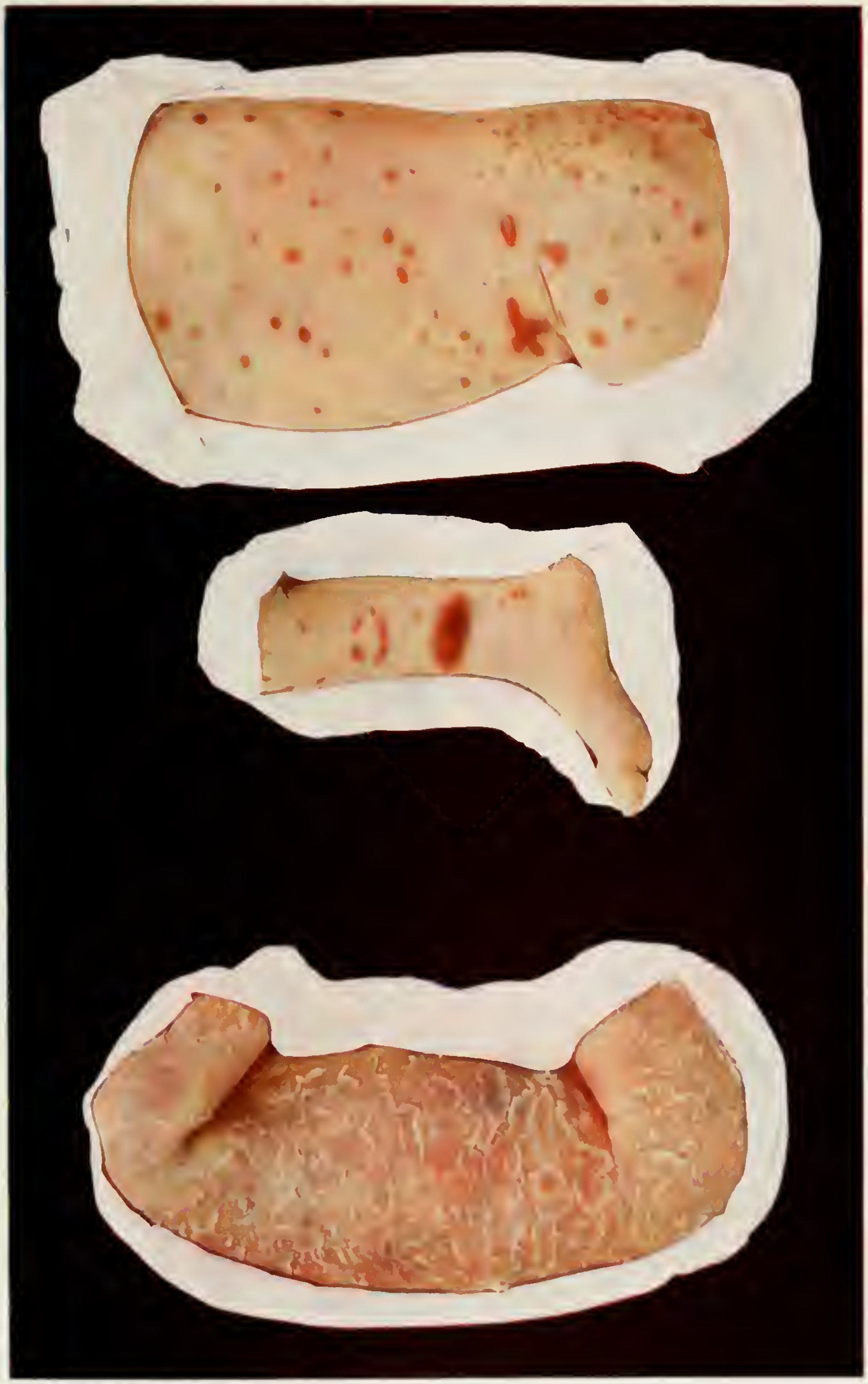
Treatment.—The treatment demands in the first place baths and in the second place medication of the skin. Therefore bran or oak bark baths should be prescribed, while powder and ointment treatment should be resorted to, to effect a healing of the affected surfaces.

DERMATITIS EXFOLIATIVA (*Ritter*)

In 1870 Ritter von Rittershayn first described this affection which had been first observed in the Foundlings Home in Prague. It is not yet established as a pathological picture, and although it probably belongs to pemphigus neonatorum, it has still to be described separately (Plate 67).

Clinical Picture.—Usually the affection commences in the second week, but has also been observed in the first and up to the fifth week. The normal desquamation of the newborn infant being completed, a considerable hyperæmia of the skin appears, beginning at the fissure of the mouth, at the lower half of the face, forming fissures at the angles of the mouth and spreading rapidly over the body. The hyperæmia becomes more intense, the skin becomes thicker and more œdematous, vesicles detach the corneal layer from the rete and can be stripped off with the finger. Extensive areas of the epidermis become detached and can be pulled off in large pieces. The buccal mucous membrane may also be involved, being covered by grayish white erosions; in severe cases the epithelium of the cornea may likewise be involved (Elliot). The course of the affection is usually afebrile, only in exceptional cases has fever been observed (Escherich). Gradually the new covering of the epidermis is formed, after which recovery takes place; otherwise the detaching process of the skin continues and a rapidly fatal termination ensues in consequence of the great loss of lymph and secondary infection.

PLATE 67.



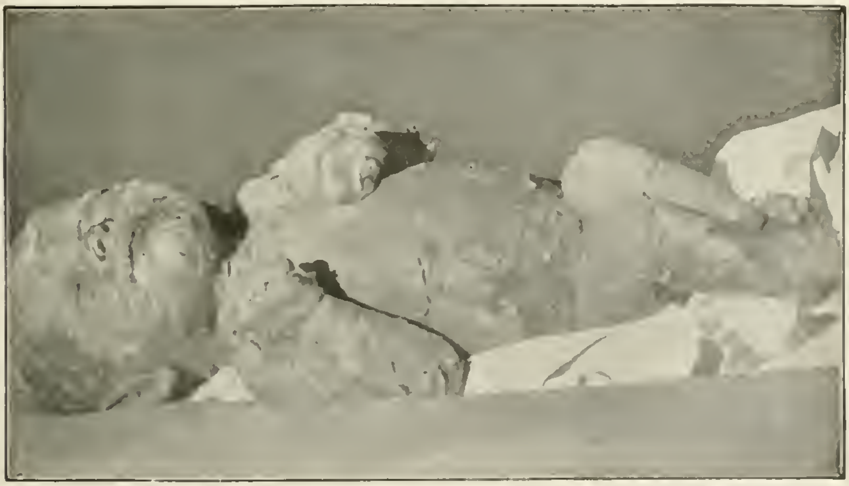
a. Dermatitis exfoliativa (light case). *b* and *c.* Urticaria pigmentosa.

The affection generally lasts one to two weeks; according to Ritter one half of the cases recover, the other half die. Intestinal catarrhs, pneumonia and ulcerous affections have been reported as complications; abscesses and furuncles may also add their quotum.

Pathology.—According to the investigations of Luithlen and Winternitz there is a dilatation of the vessels, œdema of the papillary and subpapillary tissue with considerable small-celled infiltration. Luithlen claims that there is considerable proliferation of the rete and defective cornification, while Winternitz states there is almost total absence of the same.

Etiology.—The etiology of dermatitis exfoliativa is as yet completely obscure. Ritter looks upon the disease as pyæmic, Escherich believes in a general septic infection, Luithlen in the influence of toxic factors, which seems to be supported by the appearance of the primary erythema.

Fig. 75.



Dermatitis exfoliativa. The fissures around the mouth are characteristic.

Diagnosis.—To establish the diagnosis is comparatively easy, the formation of fissures at the angles of the mouth being particularly characteristic. It is impossible to mistake these fissures for hereditary syphilis on account of the general pathologic picture.

The **prognosis** is unfavorable, the mortality being 50 per cent. These are Ritter's figures which, aside from isolated cases, are the only ones available for statistical purposes.

Treatment.—The treatment endeavors to improve the general condition and by proper bandaging to avoid injuries to the sensitive skin (packing in cottonwool). At the same time suitable baths of bran and oak bark should be prescribed to make the skin firm and promote healing. Salves and pastes (boric ointment, zinc paste, etc.) are to be recommended and limewater liniments in extensive defects.

ECTHYMA

By ecthyma we designate an affection closely related to impetigo, in which the ulcerous vesicles—contrary to impetigo—stand on a coarsely infiltrated hyperæmic base, healing sooner or later with a pigmented cicatrix.

Pathological Picture.—At first a small hard coarsely infiltrated knot of vivid red color appears; on this a small vesicle appears which soon becomes cloudy. It then ulcerates and in the course of a few days it dries up and either sinks in or grows peripherally and may attain the size of about a silver dollar. This is surrounded by a hyperæmic inflammatory areola, which persists for some time. Healing is rather protracted. The ecthyma pustules are principally at the lower extremities, the nates, rarely at the trunk. They are secondary manifestations and may be regarded as a superficial skin affection which occurs in children of any age by transmission of pus germs as a result of scratching.

Pathology.—The ecthyma pustules are distinguished from those of impetigo, by a pronounced inflammation of the cutis, which is accompanied by œdema. The papillæ are abundantly infiltrated with leucocytes. Unna looks upon ecthyma as an inflammation of the epidermis with secondary ulceration.

Etiology.—Vidal demonstrated the fact that the ecthyma pustule is inoculable; its transmission is unquestionably performed by the staphylo—and streptococci (Leloir).

Diagnosis.—For purposes of differential diagnosis there are practically only superficial ulcerous affections to be considered. In contradistinction to ecthyma the syphilitic crusts are arranged in laminations resembling oyster shells; pruritus and scratch effects are absent. The differentiation from impetigo consists in the absence of the hyperæmic indurated infiltrate in the latter affection.

The **prognosis** is benign.

The **treatment** corresponds to that of impetigo. One of the principal things to be attended to is good nutrition and local cleanliness. The best agents are sulphur baths and sulphur paste bandages.

ECTHYMA GANGRÆNOSUM

Multiple cachectic gangrene of the skin (O. Simon), pemphigus gangrænosus (Stokes), dermatitis gangrænosa infantum (Crocker), varicella gangrænosa (Hutchinson), impetigo gangrænosa (Kreibich) etc.

Ecthyma gangrænosum, in contradistinction to ecthyma, commences with the formation of indurated, dirt-colored to bluish red knots, on which vesicles promptly develop; these rapidly become peripheral ulcers which grow inward.

Pathological Picture.—The ulcers described above which are striking by reason of their sharp demarcation and appear to be cut out with an iron die, rapidly become larger and may attain the size of a pea to a five-cent piece; if close enough, they may run into each other. The base of the ulcer is hæmorrhagic and necrotic, surrounded by a hyperæmic indurated, infiltrated areola. The ecthyma pustules develop gradually, principally at the trunk (nates), extremities, and the neck. They are chiefly met with in children (tuberculosis, atrophy, etc.). If healing takes place, the necrotic portion is desquamated, forming a firm pigmented cicatrix. In the majority of cases, however, death ensues in consequence of sepsis which originates from the various gangrenous foci.

Pathology.—According to the investigations of Hitschmann and Kreibich there is a necrosis of the epidermis and corium, and a local, dense accumulation of bacteria in the infiltrated layers of the epidermis and around the vessels.

Etiology.—In view of the site at the nates we must look for the source of the infection in the soiling from urine and feces. While Baudouin and Wickham found streptococci in one case, Ehlers, Neumann and Oettinger, especially however Kreibich and Hitschmann, hold the bacillus pyocyaneus responsible for ecthyma gangrænosum. The last two investigators also consider the bacillus pyocyaneus responsible for the severe general conditions.

Diagnosis.—The differential diagnosis as against impetigo and ecthyma is established from the ulcerous desintegration and the grave course of ecthyma gangrænosum.

Prognosis.—Bad.

Treatment.—The chief object of the treatment consists in improving the general condition. Therefore the greatest importance should be attached to providing as nutritious a diet as possible (aside from iodide of iron, etc.). The local treatment should pay special attention to utmost cleanliness, the pustules should be opened, and the cleansing of the ulcers be promoted by baths (sublimate, potassium permanganate). Simultaneously bandages of iodoform and its substitutes (xeroform, airol, etc.), and salves should be prescribed for the protection of the skin.

FURUNCULOSIS

By furunculosis we understand a circumscribed inflammation caused by pus cocci situated in the subcutaneous cellular tissue, which may lead to ulceration or necrosis. According to whether the affection originates from folliculitis or the cellular tissue generally, we distinguish between furunculosis of the sebaceous glands or of the cellular tissue.

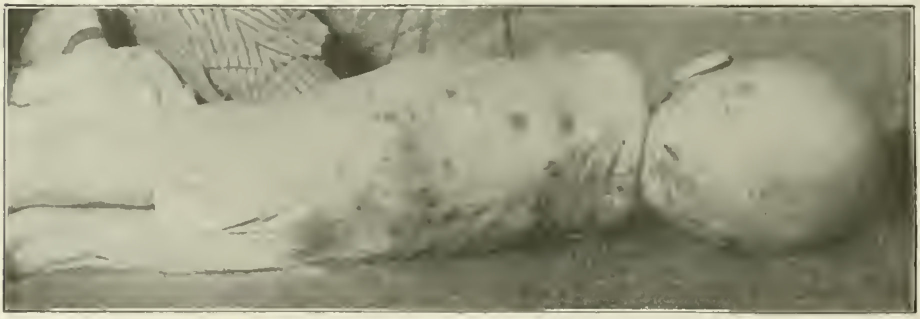
Furuncles are met with in nursing infants and in every age of childhood, originating from skin affections in consequence of itching and scratching, the ulcerating factors being transmitted through feces and urine to the macerated skin. In nursing infants they are situated principally at the upper lip, the auditory canal and the nates, and are very frequently to be found in prurigo, eczema, impetigo, etc. The course of furunculosis in children corresponds exactly to that in the adult and requires no special discussion.

Etiology.—Furuncles are caused by the penetration of staphylococci into the skin. In how far internal causes or the milk of the mother may cause infection from the intestine, has not yet been demonstrated.

The **prognosis** is favorable, provided the number of the furuncles is not excessive and the condition of the child is not too low.

Treatment.—The object of the treatment is the quickest possible evacuation of the furuncles, and keeping the skin clean. Fresh furuncles should be softened with hot poultices or plasters (salicylic soap plasters, mercurial carbolic plastermull) and then incised; afterwards apply moist bandages with acetic alumina and finally bandages of salicylic soap plaster. The cleansing of the skin is effected by frequent bathing (soap, sublimate, or sulphur baths) and regular washing with soap (sulphur soap).

Fig. 76.



Multiple skin abscesses in atrophic infant.

MULTIPLE ABSCESS OF THE SKIN

Synonyms.—Furunculosis multiplex infantum, dermatitis folliculosa (Steiner), dermatitis phlegmonosa (Baginsky), circumscribed phlegmon of the superficial fascia (Bohn).

The multiple abscesses of the skin are an affection peculiar to the nursing period; they are not connected with the follicles, but are abscesses of the superficial fascia which by reason of their slight inflammatory manifestations and their clinical course totally differ from furuncles.

Clinical Picture.—In the newborn, and as a rule in poorly nourished atrophic or tuberculous children, whose skin is flabby and atrophic, facilitating the entrance of inflammatory agents, we often see very numerous skin abscesses occurring in paroxysms singly or in groups. There may be many hundreds of them, situated at the back, neck, nates, scalp, upper arms and thighs, and also diffuse over the entire body. The skin over the abscesses is pale or highly hyperæmic, the abscesses fluctuate, finally break open or have to be incised. Their size varies between that of a pea and a hen's egg, they may even give rise to phlegmonous and gangrenous inflammations. Immediately after the incision they sink in. The pus they contain is yellow or yellowish green, and has a peculiar odor. The affection generally passes off without fever or possibly with very slight elevation of temperature, and is met with not only in impoverished nurslings, but in isolated cases also in healthy, well developed children. Occasionally complications may occur (gastro-enteritis, bronchopneumonia), but their connection has not yet been demonstrated.

Etiology.—Without doubt the abscesses are caused by microorganisms, in all probability by the staphylococcus pyogenes aureus. Renault found it fifty times in fifty cases, Hulot ten times in ten cases. Aside from this coccus, streptococci have been demonstrated and also coli bacilli in the region of the anus. Whether the microörganisms come from without or whether there is a possibility of an internal infection (hematogenously, by the intestinal tract, through the milk?) is as yet an open question. Even Escherich's opinion that the infection was transmitted by the sweat-glands does not seem to cover all cases.

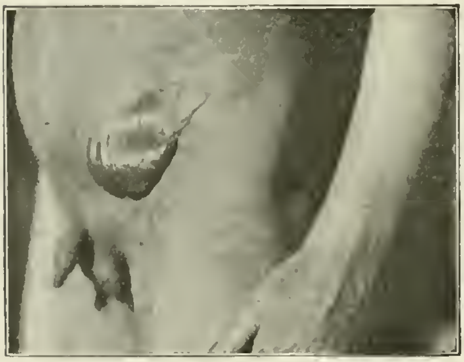
Prognosis.—The prognosis generally is good under sufficient energy and attention, provided the children are not too poorly nourished and have not suffered too long from the abscesses. In severe cases, when the affection becomes chronic and fresh paroxysms continue to occur, the patients finally succumb to sepsis or some complication in consequence of nutritive disorders.

Treatment.—The treatment has to pay attention to the regulation of the diet and nutrition so as to improve the general condition and to prevent the spreading of the ulceration by keeping the skin clean, especially by washing with sulphur soap and giving regular sulphur baths (the best are made with 30–50 Gm. of Vlemingkx' solution). The regular change of the body linen is of special importance. The furuncles should be opened as early as possible, and as many as possible every day. After the incision which should be as small as may be consistent, the child should be immediately put into a bath. Neisser makes the incision while the child is in the bath and has been very successful with this method. The healing of the abscesses should be accelerated by sulphur ointments and sulphur pastes, while the skin and the neighborhood of the abscesses should be kept clean by washing with spirits of benzine.

EPIDERMOLYSIS BULLOSA HEREDITARIA

Clinical Picture.—The following is the typical pathologic picture as it was first described by Goldscheider in 1882 and later by Köbner: Suddenly, often in summer, there occur vesicles on the normal skin at all places of the body from a minimal external cause (friction, pressure, etc.) without inflammation. The fluid they contain is generally limpid, only rarely blood stained serum. The affection attacks particularly the feet, calves, hands, but may occur at any part of the body. The disposition to this cystic formation is congenital and has been observed through several generations (through four and five by Valentin and Bonaiuti). The affection occurs chiefly in summer and especially when the skin has been exposed to some irritation. Should the feet be affected, the complaints may be so severe that the children are unable to walk. The affection strangely attacks almost exclusively children of German descent, a fact for which sufficient explanation has not as yet been forthcoming. The cysts heal without leaving any scar worth mentioning; a pigmentation only rarely remains. The affection may persist for life and cause considerable trouble. Aside from the typical form, a series of authors (Darier, Hallopeau, Fox, etc.) have described a second group, which materially differs from the first. This frequently shows keloid cicatrices, miliary cysts, very pronounced pigmentation, trophic disturbances, frequent involvement of the mucous membranes, nail affections, etc. (Hallopeau: Forme bulleuse et dystrophique). Whether this second form really belongs here or not is still a matter of contention. Vidal and Hallopeau have even described a third form (la forme fruste), in which the cystic formations are of comparatively less importance, or rather exist only in infancy, and in which instead the atrophic changes of the skin play the principal rôle.

Fig. 77.



Epidermolysis bullosa hereditaria.

The **etiology** of the affection is unknown and heredity plays a most important part. Thus Valentin observed 11 members of one family suffering from it in four generations, Bonaiuti saw 31 afflicted in a family of 63 members, Bettmann saw 10 cases among the offspring from one grandmother.

The **prognosis** in spite of chronicity in isolated cases, is relatively favorable, whereas the cases which are accompanied with trophoneurotic disturbances are distinctly less favorable.

Treatment.—Treatment is comparatively helpless. In one case Jarisch observed a favorable effect from arsenic, otherwise the treatment can only be directed toward the protection of the skin and the alleviation of the subjective complaints.

HERPES

By herpes we understand the sudden appearance of small grouped nodules upon a hyperæmic skin, rapidly changing into cysts and having the appearance of vesicular eruptions. There is generally no fever, the vesicles exist only for a short time and within 24 or 48 hours the fluid they contain dries up, the covering skin turns into a scab, and in a few days healing may be complete.

Clinical Picture.—Herpes in children is principally situated in the vicinity of the mouth, lips, nasal apertures and cheeks, in the form of herpes facialis. In children who are specially predisposed, it occurs relatively often, either following certain infectious diseases or without any assignable cause.

The **prognosis** is distinctly favorable, the vesicles healing in a short time. For purposes of differential diagnosis herpes zoster (see next paragraph), varicellæ (diffusion over the entire body) and hydro-vacciniforme, which occurs under the influence of light, have to be considered.

The **treatment** consists in protecting the vesicles, that is, in keeping the cystic cover intact in order to prevent the penetration of infectious agents. Here a simple salve, cooling ointment or paste is indicated; a very good effect is exercised by Unna's unguent. caseini (P.G.) which, when rubbed into the skin with a trace of water, forms a protective covering over the vesicles.

HERPES ZOSTER

Herpes zoster (Gürtelrose, belt-rose) is an affection which often appears accompanied with light febrile manifestations and is characterized by herpetic vesicles on hyperæmic ground which are arranged according to some system.

Clinical Picture.—In infancy, and even in the earliest days of life, under slight prodromal manifestations (elevation of temperature, itching and burning of the skin), eruptions of groups of small nodules occur which change into vesicles and generally follow the track of a nerve. The nodules or vesicles usually appear in the region of an intercostal nerve (hence the name belt-rose), there are spasmodic recurrences for a few days until the median line has been reached; only rarely is the latter over-reached either in front or behind. Usually the eruptions remain unilateral. According to its appearance we distinguish herpes pectoralis, herpes facialis, herpes frontalis, etc. The shoulders and extremities are but rarely involved in children.

Herpes zoster is a comparatively infrequent affection among children in Germany. Comby counts one case in a thousand; personally I have only seen two cases in my children's policlinic. It is very rare under two years of age, rarer still under four years, after that it occurs more frequently. It is a surprising fact that twice as many girls are affected as boys. On the other hand Bateman has found a preponderance of young people between twelve and fifteen years suffering from this complaint. Crocker found 75 per cent. under twenty years. Evans observed that half his cases of herpes zoster occurred in children under fourteen years. Whence this frequency arises (race, climate, etc.), is not yet cleared up.

The complaints in children are the slighter, the younger the child. It is only after they have reached the age of ten (Comby) that the pain becomes more pronounced. Neuralgia, so trying in the adult, seems to be entirely absent. On the other hand there is often slight glandular enlargement and light keratitis (Millon). Besides, photophobia (von Gewaert) and facial paralysis (von Epstein) have been described. The fever rarely exceeds 38° C. (100° F.) maximum 40° C. (104° F.) and lasts no longer than five to six days.

Pathology.—The origin is unknown. We know that shingles appear epidemically (fall and spring), also that toxic substances (arsenic, carbon oxide gas, pyæmic processes, typhoid, etc.) may produce herpes zoster. Central causes (injuries to the gray substance) may occasion zoster (Head, Babes, etc.). According to Barensprung nervous disturbances may be the cause which in their turn are occasioned by the penetration of infectious agents into the peripheral nerve terminations or into the intervertebral and spinal ganglia.

In how far family transmission (Klamann, Millon) or local tuberculosis may be responsible for zoster, is still subject to demonstration.

The **diagnosis** is easy, and the **prognosis** favorable, as the cases run a light course and relapses in children are infrequent.

Treatment.—The treatment consists, similarly to simple herpes, in the protection of the skin and the preservation of the cystic cover by means of salves, pastes and bandages. In these cases, too, Unna's unguent. caseini (P.G.) is of value.

SYMMETRICAL GANGRENE (*Raynaud*)

Symmetrical or Raynaud's gangrene is characterized by localized ischæmia which always occurs in paroxysms, by its symmetry and the disturbance of circulation (local asphyxia), regional cyanosis, which later may lead to necrosis. The affection, which is very rare in childhood, generally attacks the fingers (or hands), toes (or feet), ears and nose. It is said to occur after infectious diseases (scarlet fever, measles, etc.) and after exhaustion. Gaspardi observed symmetrical gangrene in a three-year-old child, Behrend in one of six years, Durando Durante in two cases of newborn infants with fatal termination, whose parents were syphilitic.

The **differential diagnosis** in the case of children has to confine itself to congelation and scleroderma; in both the paroxysmal, spasmodical attacks are absent.

Treatment is comparatively powerless. Its object is to improve the general condition and to treat the local vesicular affection by electricity. If necessary, surgical interference has to be resorted to.

ATROPHIC INFLAMMATIONS OF THE SKIN (ULERYTHEMA)

Among the atrophying inflammations of the skin, that is, circumscribed inflammations which later lead to atrophy, lupus erythematodes (ulerythema centrifugum, Unna, an erythema which leaves a scar), occurs very rarely in childhood. Ulerythema ophryogenes is found more frequently in comparison. The affection which Tanzer described in 1889, commences in early life, especially in children with blonde hair, attacks chiefly the eyebrows and may spread to forehead, cheeks, neck, the extensor surface of the upper arm and the hairy part of the head. At first there is hyperæmia of the skin, and small horny cones are formed in the place of the hair follicles. The hair is abnormally thin or invisible or broken off. The erythema leads to follicular and intrafollicular atrophy and to the formation of small scarlike depressions.

Etiology.—Unknown.

Treatment.—Soaps and salves of sulphurated salicylic tar.

PSORIASIS

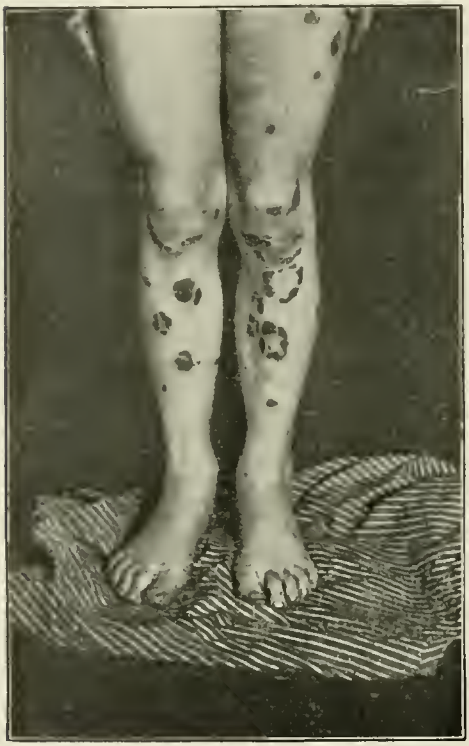
Psoriasis is a chronic affection of the skin with frequent relapses, characterized by light red papules or plaques which are covered with silvery scales. The scales are loosely connected and become easily detached through scratching, and underneath the scales appears the inflamed, light red, punctiform, bleeding skin.

Clinical Picture.—Psoriasis forms roundish patches, from a pinhead to a dollar piece in size, which gradually grow larger and may become confluent. They never exudate, itch but rarely, exhibit only slight inflammation and are covered with silvery scales. According to the size or the confluent character of the patches we distinguish psoriasis punctata, guttata, annularis, gyrata, etc.

Psoriasis attacks with great predilection the extensor surfaces of the extremities, the hairy part of the head and the skin over the sacral bone; the nails also may become fissured and brittle. The affection is characterized by the absence of pruritus and other symptoms, by the presence of silvery scales and by its chronic course. It recurs in spasmodic paroxysms and may last through life. According to

Nielsen in 44 per cent. of all cases it commences before the fifteenth year, generally after the fifth or sixth. It may, however, also appear in earliest infancy (on the 38th day, Rille; in the fourth month, Neumann). In children it runs a relatively benign course; severe cases have not been as yet reported. There is a total absence of complaints, and only as fresh relapses occur, there is sometimes considerable pruritus. It is often very strongly developed on the hairy part of the head in childhood, thick scaly masses being formed there.

Fig. 78.



Psoriasis in a young girl. (Psoriasis gyrata et annularis). Typical localization at the knees.

Pathological Anatomy.—The microscopic picture shows a broadening of the stratum corneum and a thickening of the rete Malpighii with considerable proliferation. The papillæ are enlarged, swollen and œdematous, and there is considerable dilatation of the papillary vessels. The vessels of the corium are dilated, with slight small-celled infiltration which quite corresponds to the clinical picture of absence of coarseness in the psoriatic plaques.

The **etiology** of psoriasis is entirely unknown (diathesis? neuropathic etiology? mycotic causes?). Quite a series of cases that occurred in childhood have been published, in which psoriasis has appeared following auricular eczema and piercing of the lobe (Henoch, Neumann). Whether the occurrence of psoriasis after vaccination has any connection with the latter, is not proved. Nor do we know whether heredity has anything to do with it, whether in family transmissions it is congenital, whether the tendency is inherited, or whether there is a simple infection.

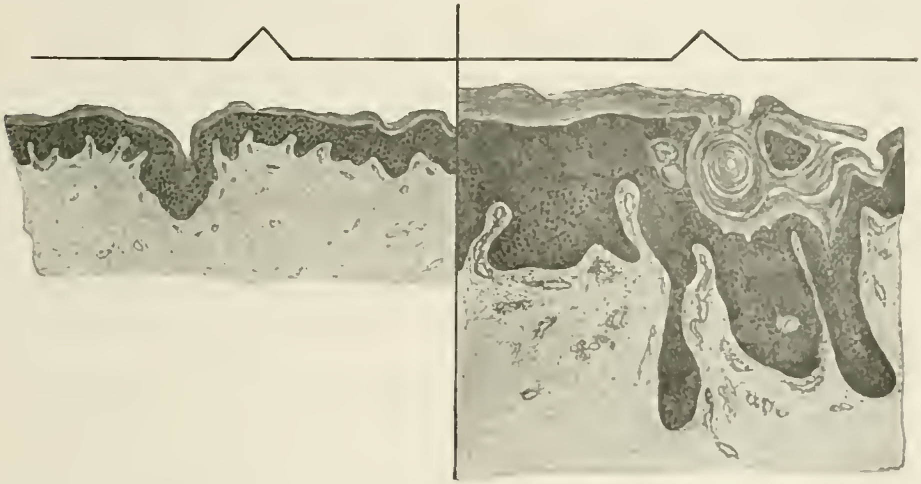
Prognosis.—The fact that psoriasis is a chronic affection which is difficult to cure does not make the prognosis very favorable. Even though it causes no trouble during childhood, it should be remembered that an afflicted child may never lose the eruption for the rest of its life.

Diagnosis.—The diagnosis of the light form occurring in childhood is much easier than in the adult, as the silvery scales and the bleeding spots which occur after scratching them off, excludes any possible confusion.

Treatment.—The object of the treatment is to influence the eruption by internal medication, to remove the scales by external measures and to effect a healing of the exposed surfaces by medicinal agents.

Of *internal medicines* only arsenic is to be considered. In younger children it is administered as Fowler's solution 2.2 Gm. (½ dr.) aq. destill. 8.0 c.c. (2 dr.) (5 gtt. t.i.d.), or in the form of injections (liquor pot. arsenicosi, aqua destill. aa 5.0 c.c. (1½ dr.) 1 unit of the Pravatz syringe up to 15 units, according to age), in older children, when anæmia is present, in the form of the well-known pills of iron and arsenic. Generally speaking, the arsenic treatment is not reliable; still more uncertain is the efficacy of the substitutes sodium cacodylate and atoxyl. Under certain circumstances the diet has to be changed, an improvement being frequently effected through a change in nutrition.

FIG. 79.

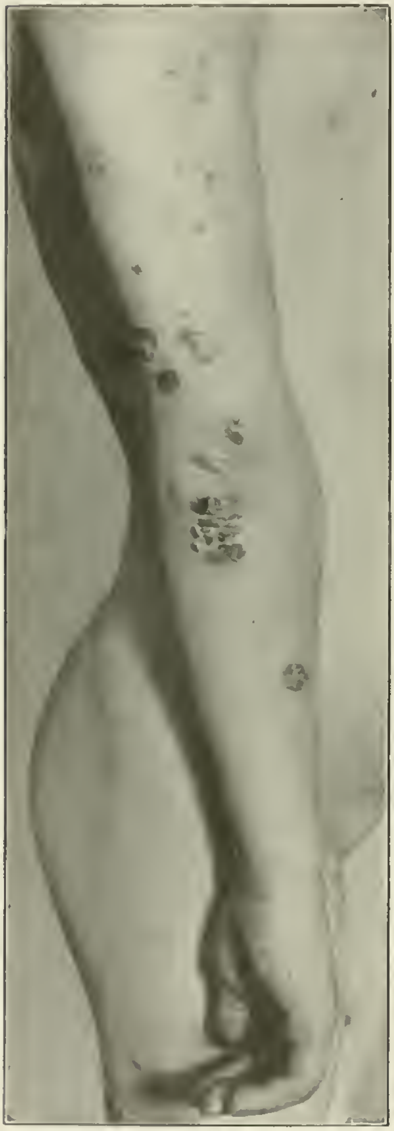


Psoriatic skin contrasted with normal skin (left). Expansion and thickening of the rete with considerable proliferation, enlargement of the papillæ with small-celled infiltration.

The *external treatment* consists in bathing, sweating, application of soaps and plasters, removal of the scales by rubbing, and softening of the psoriatic plaques, in order to prepare the skin for the real medicinal treatment. Children are given daily warm baths 28°–30° C. (82–86° F.) lasting half an hour to an hour, applying either sulphur soap or green soap. Simultaneously the scalp is anointed with tincture of soap and cleansed in the bath. Sulphur may be added to the bath, or immediately after the soap treatment in the bath tar may be applied (see Tar Baths p. 434). As soon as the child has left the bath, the body is treated with medicaments, that is, larger surfaces are anointed and bandaged, smaller ones and localized foci are painted and a softening plaster applied over the coat (zinc oxide plastermull, salicylic soap plaster). The best and mildest ointment for the skin, especially scalp and face, is a combination of ung. hydrargyri album and tar. I prescribe the ointment as follows:

hydrag. precip. alb. 2.0–4.0 Gm. (½–1 dr.) bism. subnit. 4.0 Gm. (1 dr.) anthrasol 0.5–2.0 Gm. (7½–30 gr.) ung. leniens ad 40.0 Gm. (1¼ oz.). This ointment may be made more liquid by an addition of olive oil for rubbing into the scalp, especially in the case of girls. Salicyl 0.1 Gm. (1½ gr.) may be added to soften the scales. The best ointment for application on the body, chrysarobin, is now generally introduced and prescribed for children as a 1 per cent. ointment; for painting 1:100 traumacine. (Caution on account of serious dermatitis and conjunctivitis). Kromayer recommends eurobin as a substitute for chrysarobin which is prescribed together with eugallol (substitute for pyrogallus) as follows:—eurobin 1.0 Gm. (15 gr.) eugallol 1.0 Gm. (15 gr.) acetone ad. 50.0 Gm. (1½ oz.) for painting the affected places. Pyrogallus too should be used with caution in children, as it may cause poisoning. It is therefore advisable in children to apply a weak ½ to 1 per cent. ointment of pyrogallus. As above mentioned, plasters are now applied over the painted places; or instead of painting, the chrysarobin plastermull, which sometimes has a strongly irritating effect, may be applied. Generally speaking, the chrysarobin and pyrogallus preparations cannot be handled carefully enough, the infant's skin being exceedingly sensitive. A very good effect is obtained by tar baths, which should be given several times a week, after which the affected places should be bandaged with chrysarobin or pyrogallus. Also painting with tar, the colorless liquor carbonis detergens or the more powerful tincture of tar (pix liqu. 3 per cent. alc. ad 30.0) can be recommended, but should also be used with caution. In carrying these measures out, it is advisable to make regular examinations of the urine. As mild agents may be mentioned sulphur baths and sulphur pomatum (1–5

Fig. 80.



Psoriasis in young girl. Typical localization at the extensor surfaces of upper and lower arm.

per cent.) mixed with ½ per cent. anthrasol. For the treatment of the hairy part of the head precipitated bismuth tar ointment and salicylic oil (salicyl 10 Gm., ol. ricini 40 Gm., ol. oliv. 50 Gm.) may be used with or without anthrasol; for the nails, scalp treatment, baths and plasters.

SCLERODERMA

Scleroderma is a chronic affection of the skin which usually commences with œdematous swelling, leading to a coarse thickening and induration of the connective tissue of the skin and ending in atrophy.

According to the extent of the scleroderma we distinguish the universal forms (sclérémies, Besnier) and the circumscribed forms (morphœa). Generally speaking, three forms may be differentiated: (1) diffusely spread over the entire body, commencing with coarse, glistening, œdematous swelling, gradually leading to a stone-hard induration, and often healing rapidly (sclérodermie œdemateuse, Hardy); (2) the form commencing symmetrically in the face, at the head and extremities, gradually becoming diffuse, often leading to sclerodactylia (isolated involvement of the hands with trophic disturbances of the muscles, vessels and bones) and designated sclérodermie progressive; (3) the localized forms which are sometimes called sclérodermie en plaques (morphœa, Wilson) and sometimes sclérodermie en bandes, according to whether the eruption is arranged in roundish foci or in long stripes. In this form the freshly inflamed zone is frequently observed as a bluish ring, glistening with a violet tint, about 2 to 10 mm. wide (lilac ring of the English), which encircles the coarsely infiltrated, atrophic yellowish or brownish centre.

The affection often commences with prodromal manifestations (sensation of cold, pain, itching etc.). There is also erythema or œdema at the attacked places, the skin becomes hard, rigid, difficult to lay in folds, looks bluish white, arrests the movements, so that the face assumes a peculiar mask-like expression, while the fingers acquire that immobility and rigidity which is known under the name of sclerodactylia. Gradually the affection leads to a thickening of the skin and atrophy, but it may also undergo spontaneous cure. Other signs are reduced sensibility and sweat secretion, falling out of the hair, accompanied by increased pigmentation; facial hemiatrophy has been described by Neisser and Jagot.

The affection is found comparatively rarely in childhood, but may occur in the first few months of life (even in the second and third weeks—Cruse, Silbermann, Baldoni, Neumann). It attacks both boys and girls without distinction. Haushalter observed sclerodactylia in a seven-year-old child.

Pathology.—The chief nature of the disease is an affection of the connective tissue which is changed to a swollen, homogeneous, glassy layer, appearing saturated with a viscous fluid which has coagulated (Neisser). The elastic fibres are reduced, the vessels are partly obliterated, partly stenosed.

Etiology.—The cause of scleroderma is unknown. It is either a trophoneurosis or a nutritive disturbance of the connective tissue and vessels.

Prognosis.—The prognosis is relatively favorable in childhood, the affection running a lighter course than in adults; in the nursing period the affected places nearly always heal.

Diagnosis.—Recognition of scleroderma is comparatively easy from the induration, coarseness and rigidity of the skin.

Treatment.—Sternthal's dictum that "scleroderma either heals spontaneously or not at all" is certainly not justified in view of the various methods of treatment by which the disease can be favorably influenced. By internal treatment and invigorating diet the general condition should be fortified, and at the same time arsenic, quinine and strychnine administered, the best method being the three together in pills. Thyroidin has also been recommended. By local treatment the skin should be softened and the affected parts rendered more mobile. This is best accomplished by hot water and hot sand baths, hot douches, steaming, sweat cures and moist packs. The medicines to be most recommended are salicylic preparations in connection with soap and salicylic soap plaster. In recent times experiments have been made to soften the skin, which is as hard as a board, by the application of thiosinamin (Hebra, Neisser, Galewsky). Every one or two days ¼ to 1 c. c. of this substance with a 10 per cent. solution of glycerin is injected in the neighborhood of the diseased skin, or directly applied to the affected places in the shape of thiosinamin plastermull. I have been very successful in this way in three cases. At the same time massage and electric treatment (constant current or electric baths) are particularly valuable. Brocq recommends electrolysis for the treatment of the circumscribed forms.

XERODERMA PIGMENTOSUM (*Kaposi*)

Under the name of Xeroderma pigmentosum, Kaposi described in 1870 a chronic affection of the uncovered parts of the skin occurring under the influence of the rays of the sun, which leads to pigmentation and atrophy and is accompanied by the formation of malignant tumors.

Clinical Picture.—Small brownish maculæ resembling freckles, appear on the skin, sometimes in the first year of life, which make the skin look spotted. In the interstices there are dilatation of the vessels, small verrucous formations, papillary and scaly deposits. The skin between these spots looks white and atrophic. Gradually the affection spreads, the pigmentation of the skin increases, and the skin becomes more and more atrophic. There is xerosis of the conjunctiva, ectropium, ulcers and fissures of the skin, and multiple pigmented sarcomata and carcinomata may appear even in the third year.

The affection occurs in earliest life (first and second year) and is doubtless attributable to a family disposition. Although parents and children are never affected simultaneously and although heredity can never be demonstrated, the influence of race and family disposition is unquestionable. According to Halle the disease seized 88 families in 186 cases, Forster found 150 positive cases distributed among 96 families. Besides there was consanguinity of 11.5 per cent. as against the average frequency of 6 to 11 per cent. in marriages of relatives. It is remarkable that usually the children of the same sex are affected in a family. Jewish children seem to be specially prone to this affection, Elsenberg observing 24 per cent. of Jews in 52 cases. The affection is in all probability caused by a hypersensitiveness to light, as it usually occurs at the uncovered parts of the body, the face, hands and, if exposed, the feet.

Pathology.—Microscopic examination of the first stage shows an accumulation of round cells in the papillary body and stratum subpapillare (Lucasiewitz), and a distinct œdema around the blood vessels and glands. In the later stages there is atrophy of the tissue in the white atrophic places, while in the hyper-pigmented places there is a thickening of the papillary body, and in it cellular infiltrates together with considerable accumulation of pigment. The vessels, especially the capillaries, are considerably changed.

Etiology.—As to the cause of the affection itself and the development of sarcoma and carcinoma, we are as yet completely in the dark.

Prognosis.—Unfavorable.

The **diagnosis** does not offer any difficulties.

Treatment.—The treatment is powerless to remove the cause; perhaps it is possible to eliminate the influence of the light by protecting the skin through colored veils, etc., perhaps it is also possible to effect a cure of the carcinoma and sarcoma by phototherapy.

ANOMALIES OF THE SWEAT SECRETION

HYPERIDROSIS

Excessive sweat secretion may be universal (hyperidrosis) or local (epidrosis). Quite a number of children, especially nervous ones, react to every irritation by perspiration. On the other hand excessive sweating may also be caused by heat, the sun, clothing, etc.

Excessive local hyperidrosis is found on the scalp during the first few years of life, in later years it is localized, particularly at the hand and feet. It is frequently met with in nervous, chlorotic children in families with a nervous taint, in anæmic girls at puberty, and as a consequence of disturbed circulation (pressure of shoes, garters, etc.). When the hands or feet experience a sensation of great cold, there occurs a partial hyperidrosis in which the sweat becomes easily decomposed, giving off an offensive odor and leading to maceration of the skin. Very often a cold perspiring foot of this description causes chilblains.

Treatment.—The first task is to treat the general causes, chlorosis and anæmia, and to influence through diet and general directions the nervous condition. At the same time all disturbances of circulation (narrow shoes, garters, etc.) should be removed. By the local treatment the excessive secretion should be reduced, antihydrotic measures being indicated, for instance baths of oak bark and nut leaves, painting the feet with chromic acid (5 per cent., caution!), washing with a 2–10 per cent. formalin solution (caution: eczema), and washing with spirit of naphthol 5.0 Gm., glycerin 10.0 Gm., spir. colon. 30.0 Gm., diluted spirit ad 150.0 Gm., or with a 5 per cent. solution of tannin. Besides, weak formalin ointments or Hebra's ointment can be recommended to apply to the hands. It is of special importance to frequently change the stockings and to keep the feet dry by dusting them with dermatol powder, if they are at all involved.

Note.—Reduction of the sweat secretion (anidrosis) and partial absence of sweat secretion occurs from nervous and psychic causes. Among the qualitative disturbances brief mention may be made of bromidosis (unpleasant odor of the perspiration) and chromidosis (changed color of the perspiration) as being connected with hyperidrosis. Chabbert communicated three cases of yellow chromidosis in the children of one family.

DYSIDROSIS (*Tilbury Fox*)

(Cheiropompholyx, Hutchinson)

Under this name Tilbury Fox described in 1871 an affection which is characterized by the acute appearance on normal skin of small vesicles, the size of sago, which contain a clear fluid. It chiefly attacks the lateral surfaces of the fingers in paroxysms, but may also involve the palms and soles. The affection may be accompanied by itching and burning, is always connected with hyperidrosis and may also produce larger cysts. It heals in one or two weeks, but is greatly liable to return.

The **etiology** is unknown, but a nervous family disposition seems again to play the principal part.

The **pathology** shows a cystic formation in the corneal layer, but a connection with the excretory ducts of the sweat-glands is not demonstrable.

Treatment.—The treatment is the use of arsenic and general directions in order to overcome the nervousness and hyperidrosis; locally formalin soap and a 1 per cent. formalin solution, also washing with spirits of naphthol, resorcin or thymol, gradually rising from ¼ per cent., are to be recommended. Should the dysidrosis be complicated by a secondary eczema, the latter will have to be removed first.

MILIARIA (*Sudamina*)

Following febrile diseases, in consequence of keeping infants excessively warm, under the influence of the sun, small transparent vesicles frequently appear, especially on the chest, which may be interspersed with small papules. According to whether these papules, which are frequently very numerous and the size of a pinhead, are white or red, we speak of miliaria alba or miliaria rubra. The minute transparent vesicles between them (miliaria crystallina) contain a limpid fluid with acid reaction. Miliaria is often itching and heals in twenty-four to thirty-six hours with slight desquamation. No doubt this is a case of obstruction of the excretory ducts of the sweat-glands and its occurrence is often regarded as a critical phenomenon. Independently of these pathologic conditions I have seen numerous cases of miliaria arise in very hot summers under the influence of the heat.

Treatment.—Treatment may confine itself to bathing, washing with water and vinegar or ¼ per cent. spirit of thymol, and dusting with powder in order to accelerate the healing process.

ANOMALIES OF THE SEBACEOUS GLANDS

SEBORRHŒA

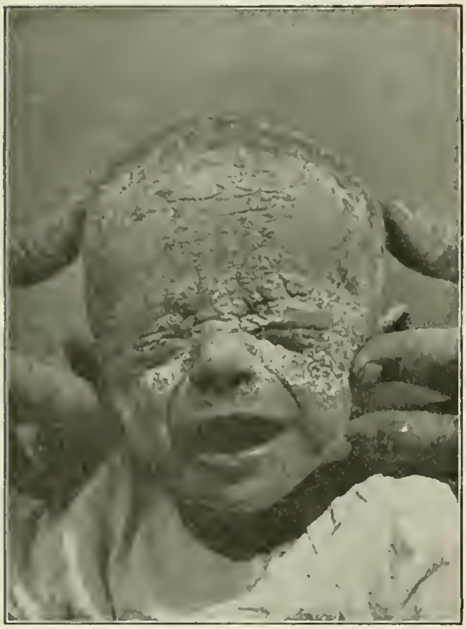
By seborrhœa is understood a pathological increase of the secretion of the sebaceous glands. It is divided into (1) seborrhœa oleosa and (2) seborrhœa sicca (furfuracea-pityroides, pityriasis capitis) according to whether the secretion of the sebaceous glands is oily or dry.

Seborrhœa oleosa is principally met with in the later stages of infancy in the shape of lardaceous, oily secretions, so that the face, especially the nose and the forehead, but also the chest, are covered with a glistening fatty layer. This is quite a harmless condition, the only treatment necessary being dusting with powder.

The second form, *seborrhœa sicca*, occurs in the first few months of life, especially on the scalp (over the large fontanelles). The scalp is covered with yellow or yellowish brown, lardaceous, scaly masses which frequently assume a gray or grayish black color owing to accumulation of dirt and dust. On removal of the scales the exposed skin looks slightly hyperæmic and macerated and is covered with a thin, oily layer. The affection may spread to the eyebrows, forehead, nose and chin, from there to the back, chest and folds of the body. All these parts are covered with yellowish scales. In the course of the first few years the seborrhœa gradually disappears, or else it may be complicated by eczema (eczema seborrhoicum).

In the later periods of childhood seborrhœa occurs in the shape of a grayish white diffuse furfuraceous desquamation, especially on the head (pityriasis capitis); the skin is covered with small whitish scales, has a milky appearance, and frequently there is a secondary falling out of the hair at this early age. This stage is chiefly found at puberty as status seborrhoicus and is occasioned (1) by an engorgement of the sebum in the glands; (2) by a cornification anomaly which leads to an occlusion of the sebaceous glands, while (3) at this period an increased production of sebum and the growth of hair act as irritations. Very often there are also seborrhœic, sharply demarcated yellowish spots covered with small scales, which are more or less distinct, and which sometimes disappear and reappear, spread to the face, the nasolabial folds, and chin, cheeks and finally to the chest, where they attack the sternum (Eczema seborrhoicum, Unna, see p. 475).

Fig. 81.



Seborrhœa sicca in nursing infant (high grade).

Pathology.—The microscopic examinations conducted principally by Pohl-Pinkus, Piffart, Unna, Ehrmann, disclose an enlargement of the sebaceous glands and a thickening of the corneal layer. In the enlarged follicles and infundibula of the hair there are broken off bits of hair and a cone called by Sabouraud "Bakterienkokon" or utriculus, which is formed of corneal cells and is pervaded by rods and microbacilli.

Etiology.—The cause of seborrhœa is unknown. Unna, Sabouraud and others regard the above mentioned microbacillus as the causative factor of seborrhœa. There is no doubt that heredity plays a rôle and that anæmia and chlorosis as well as irritations occurring at puberty in the region of the sebaceous glands form a suitable culture ground for the accumulation of organisms. Probably the disease is of parasitic origin, although proof for this assumption is yet to be forthcoming.

Treatment.—The object of the treatment is to remove the deposits and prevent the accumulation of parasites by antiphlogistic measures. We should therefore endeavor to cleanse the scalp with weak alkalies (bicarbonate of soda), with tincture of green soap, or sulphurated resorcin soap, and to soften the scales by the application of 5–10 per cent. salicylate oil, which requires an addition of 40 per cent. castor oil to dissolve the salicyl, or of salicylic sulphur ointment [ac. salicyl, 0.2 Gm. (3 gr.), precip. sulphur 5.0 Gm. (1½ dr.) ung. leniens, ol. oliv. aa 20.0 Gm. (6 dr.)]. At the same time massage, washing with sublimate (Lassar) or spirits of salicyl or resorcin, may produce an irritation of the skin which may lead to an improvement of the circulation.

ACNE VULGARIS

By comedo we designate a core which is formed in the excretory ducts of the sebaceous glands from cornified epithelial cells, sebum and bacteria. Should these comedones become ulcerative, acne pustules develop, which lead to the formation of smaller or larger, more or less hyperæmic papules around the hair follicles chiefly in the face (forehead, cheeks, chin, nose), on the chest and back as high as the scapula. These papules are either hard and red, or they may be in the ulcerative stage, or in the process of healing, the patient displaying all stages on account of the continuous occurrence of fresh outbreaks. The affection generally commences at the period of puberty and may continue for many years; it is sometimes accompanied by an anæmic or chlorotic general condition, or else by a bloated hyperæmic skin tending to urticaria which reacts to each irritation with an urticarial hyperæmia.

Pathological Anatomy.—This affection consists in an inflammatory infiltration in the perifollicular tissue, an ulcerous combination in and around the follicles with the microörganisms already referred to, various staphylococci and the acarus folliculorum. What part this parasite plays in the development of the comedones, is as yet unknown.

Etiology.—Causes for acne are furnished by the seborrhœa state (status seborrhoicus) (see page 498), hyperæmia during menstruation, anæmia and chlorosis, intestinal irritations (autointoxication) and the various forms of bacteria (staphylococci, etc.).

Prognosis.—The prognosis is favorable, although the course of the affection is a protracted one.

The **diagnosis** is not difficult in view of the presence of comedones and seborrhœa.

Treatment.—Treatment should be directed in the first place to the internal causes, chlorosis, anæmia, and intestinal disorders. The following are of foremost importance: of internal remedies iron and arsenic preparations (administered either in pills or in solution; ichthyol preparations, when there is a tendency to urticarial irritation: best with iron in the form of ferri ichthyol tablets). At the same time there should be a strict milk diet; this together with baker's yeast—the point of a knife full three times a day in a wineglassful of water—has often effected improvement. Aside from the treatment of seborrhœa (*q.v.*) the comedones and pustules should be removed. This is done by opening them with a knife or by pressing them out with the comedone squeezer (Unna). The task of the local treatment is further to dissolve the fat, remove the horny masses and open the follicles. This is done by washing with hot water and soap (either the mild "Basis-Seife" or a stronger sulphur soap), washing with benzene, hot douches especially over the body) and steaming the face (Saalfeld). An excellent effect is also produced by bathing the skin with alcoholic solutions of resorcin ($\frac{1}{4}$ to $\frac{1}{2}$ per cent.), salicylic acid ($\frac{1}{2}$ to 15 per cent.) and acetic acid (1 to 3 per cent.), but in using these remedies the tender skin of young women and children should always be taken into consideration by commencing with the weakest solutions which should often be made still milder by adding 5 to 10 per cent. glycerin. Among the ointments I recommend for the treatment of acne the keratolytic, slightly desquamative sulphurated resorcin ointments. For acne of the body stronger solutions may be applied. For acne of the neck, this part should be treated with a wash containing spirits of camphor 6.0 Gm., tinct. benz. 6.0 Gm., acetic acid 3.0 Gm.: 100 alcohol. If in severe cases of acne these measures are not sufficient, desquamation cures should be instituted, that is, aside from the treatment by washes and ointments, one to three times a week, desquamation pastes should be applied in order to denude the affected parts. In children the best plan is to apply a 5–30 per cent. resorcin-zinc paste, once, twice or three times a week, to be kept on over night, for instance resorcin 9.0 Gm., zinc oxide 1.0 Gm., amylum, vaselin aa 10.0 Gm. On the intermediate days sulphurated resorcin ointments are applied. Also moist bandages of $\frac{1}{4}$ per cent. resorcin or 10 per cent. acetic alumina may be applied over night in order to soften the acne.

MILIUM (GRUTUM, ACNE MILIARIS)

Milia are white or yellowish round papules up to millet size which are superficially embedded in the normal skin and may be expressed on pricking the skin. They are simple horny cysts, situated at the eyelids, the malar bones, temples, cheeks and lips. The treatment consists in pricking the skin and expressing them.

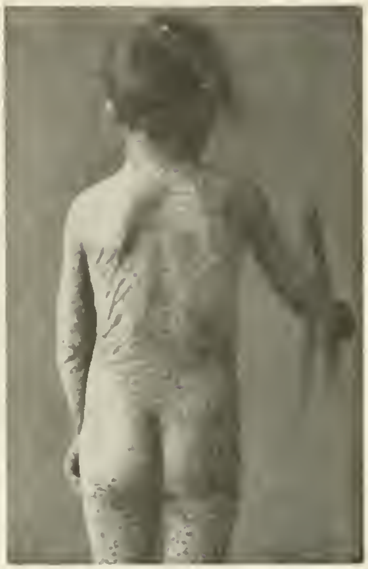
KERATOSIS (CORNIFICATION ANOMALIES)

ICHTHYOSIS CONGENITA (SEBACEA)

(Keratosis diffusa congenita)

This infantile affection was first described in 1792 and again later under a variety of different names. It is congenital and consists of an enormous deposit of sebum and corneal masses upon the skin of the newborn infant. The affection is comparatively rare (Riecke, 1901, 54 cases), and occurs at birth in weak infants. Usually the child dies in a week or sooner, although in light cases they may be kept alive for a longer period. The skin consists of large horny masses broken up by fissures and furrows, and seems too tight for the body. The mouth and its angles are distorted, the curves obliterated, ectropia and deformities of the nose develop, and the folds of the skin are absent. The disease is generally developed in utero (in the fourth month) and the child dies from inanition, being unable to suckle. In lighter cases where the covering is more parchmentlike, the attempt to keep the child alive for a longer period is successful. Riecke distinguishes between three forms: (1) true ichthyosis congenita (severe cases developed in utero; die within one to four days after birth); (2) ichthyosis congenita larvata (milder cases or those which involve the skin only partially, developed entirely or nearly so at birth; children remain alive for a longer period); (3) ichthyosis congenita tarda (birth normal, appearance of the disease at a later period; children sometimes remain alive).

Fig. 82.



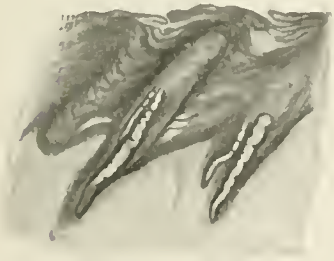
Ichthyosis serpentina or sauriasis (Crocodile Skin).

Pathological Anatomy.—There is considerable thickening of the corneal layer (up to 500 μ as against 31 to 47 μ of the normal skin, which is augmented by new formation in the rete, and rapid cornification producing diffuse keratoma. The cutis is normal (Kyber, Wassmuth, Riecke, Neumann).

Etiology.—The etiology is still completely unknown.

Prognosis.—The prognosis is generally fatal, except in light cases.

Fig. 83.



Ichthyosis serpentina. Enormously strong development of the corneal layer.

Treatment.—Baths, improvement of nutrition, embrocation with mild ointments, would seem to be the best measures.

ICHTHYOSIS

Ichthyosis is an affection which occurs in earliest infancy, usually in the first or second year, and which is peculiar for the formation of thick, adhesive scales and horny masses, also for its chronic course.

Pathological Picture.—The affection, which generally appears diffuse, rarely localized, forms more or less gray, thick stratifications upon the skin, which is dry, brittle, and divided into irregular squares which are separated by furrows. There is no itching, sensitiveness is maintained, secretion of fat and sweat diminished. The affection is very often hereditary and attacks preferably the male members of a family. Its favorite sites are the extensor surfaces of the extremities, the trunk, less often the face. The lower extremities are most frequently involved, the articular flexures are usually free; if the face is involved, it assumes a peculiar rigid expression; there is ectropium and fissures around the angles of the mouth. According to its degree of severity ichthyosis is divided into three forms: (1) ichthyosis simplex or nitida (polygonal scales, gray skin like that of a fish, described by Besnier as xeroderma or ichthyosis furfuracea, and as existing in the first few years of life); (2) ichthyosis serpentina or sauriasis, Wilson (crocodile skin, cuirass-like covering, rigid verrucous eminences); (3) ichthyosis hystrix (with special participation of papillæ, verrucous aculeate excrescences ("porcupine" beings). The course of ichthyosis is progressive up to puberty, then as a rule it stops. Eczema may occur as a complication.

Pathological Anatomy.—The corneal layer is enormously developed, the granular layer is absent, the aculeate layer is only narrowly developed. There is an immediate transition from the cells of the rete to those of the corneal layer. The papillæ are strongly developed, especially in ichthyosis hystrix. Kromeyer holds the peculiar sclerotic connective tissue responsible for the causation of ichthyosis.

Etiology.—The cause of ichthyosis is unknown to us, although there is no doubt that heredity plays an important rôle. Reyer for instance has described ichthyosis in six generations; it may pass over several generations and reappear later. It is a remarkable fact that the disease is endemic in certain regions (the Moluccas, Paraguay and the Miridites) especially with the male part of the populace.

The **prognosis** is unfavorable as far as a cure is concerned, the disease being considered incurable, although it may be relieved by treatment.

Treatment.—The object of the treatment is to remove the scales by all methods which have a keratolytic effect. There are in the first place baths: sulphur (Vlemingkx' solution 30 to 50 Gm.), potassium (50 to 100 Gm.), borax (30 to 50 Gm.), or simple soap or steam baths; also sweat cures should be resorted to for the softening of the skin. At the same time washing with sulphur soap, salicylic soap, resorcin soap, green soap are advisable, also inunction of the body with salicylic vaselin, salicylic sulphur ointment or Hebra's ointment (caution: simultaneous sulphur preparations on account of the formation of sulphuret of lead). Rheumasan ointments also have a very strong keratolytic effect. For localized ichthyosis resorcin plastermull and salicylic soap plaster are likewise recommendable.

FOLLICULAR KERATOSIS

(*a*) LICHEN PILARIS

Synonyms.—Keratosis follicularis or suprafollicularis (Unna), ichthyosis pilaris (Kaposi), xerodermie pilaire (Besneir).

Lichen pilaris is a follicular hyperkeratosis situated especially at the extensor surfaces of the upper arm and thigh. Small, acuminate, hard, horny cones rise above the level of the follicular apertures and often contain remnants of hair. The skin feels rough on palpation, in more serious cases it imparts the sensation of a grater. Often there is slight hyperæmia around the follicles (keratosis follicularis rubra). The affection is found more frequently with girls than boys; it generally commences in the second period of childhood, exacerbates until puberty and then remains unchanged. It is often met with in families and seems to be hereditary. It may persist throughout life or may heal with a follicular atrophy in the shape of small depressions.

This is a follicular hyperkeratosis with a favorable prognosis.

Treatment.—Treatment must be continued for a considerable time in order to soften the horny masses by baths, soaps and salicylic ointments.

(*b*) PITYRIASIS RUBRA PILARIS

(*Divergie-Richaud-Besnier*)

Pityriasis rubra pilaris represents a universal affection of the follicles. These are all covered with small, firm, indurated, horny cones, which are often arranged in groups (extensor surface of the phalanges!). The entire skin is hyperæmic and covered with a desquamation similar to that of pityriasis. Thick horny masses are found at the palms and soles, at the finger tips and the dorsal surfaces of the joints. The entire skin feels like a grater, looks whitish to reddish according to the stage of the hyperæmia, and scales off considerably. Desquamation and roughness continue to increase and the hyperæmia may likewise increase without any corresponding increase in the inflammatory manifestations of the skin. The affection is principally found at the antero-exterior surfaces of the extremities, in the flexure folds of the large joints, face, scalp, neck, chest, gluteal folds, phalanges of the fingers, etc. The affection is relatively rare, commences in infancy and persists for life.

Pathological Anatomy.—According to the investigations made by Jacquet and Galewsky this is a hyperkeratosis of the skin which is accompanied by a primary hyperæmia of the skin and is followed by a slight secondary inflammation of the same.

Treatment.—This is comparatively powerless and can do nothing more than relieve the feeling of tension and reduce the desquamation by softening the skin. Arsenic is without effect.

VERRUCÆ (WARTS)

By verrucæ we designate a benign circumscribed keratosis which is accompanied by an elongation of the papillæ and which is probably infectious. In children we distinguish verruca vulgaris and verruca plana juvenilis.

Verruca vulgaris is a hard, flat or raised, coarse eminence of the skin which slowly grows larger and has a rough and ragged surface; it is roundish in shape and generally of a somewhat grayish color. The ordinary wart may occur singly or in groups at the hands, fingers and scalp; very often only one wart appears first and after some time several new ones may appear simultaneously (verrue mère et verrues filles).

Verrucæ planæ juveniles are round or polygonal flat eminences from pinhead to lentil size, which grow either singly or occasionally become confluent, have a yellowish or brownish color and in contrast to ordinary warts can be easily scratched off. They are usually situated in the face and on the hands and are sometimes present in exceedingly large quantities.

Pathological Anatomy.—Marked hyperkeratosis and hyperplasia of the rete, elongation and broadening of the cones, elongation of the papillæ.

Etiology.—Through the work of Variot, Licht, and Jadassohn, the transmissibility of warts from one skin to another has been demonstrated. The length of the incubation period lasts from 5 weeks to 8 months, and this surprising length of time is probably the reason why the demonstration of their transmissibility has been so long retarded, although popular belief has always asserted the same.

Treatment.—Arsenic may cause warts to disappear, especially the juvenile form. In a number of my cases this treatment has been thoroughly successful, in others it was a complete failure, a fact foi which I am unable to account. Foulard has seen successful treatment with 1 Gm. (15 grains) magnesia per day, while others author praise the efficacy of tincture of Thuja (P.G.) (10 to 50 drops gradually increasing; caution! danger of poisoning!).

For local treatment, especially of single ordinary warts trichloride acetic acid or 10 per cent. subl. collodium, or galvanocautery may be applied, unless it is thought preferable to treat the warts by electrolysis. (Needle negative pole, piercing with the same in all directions, current 1 to 5 M.A., duration of each perforation 1 to 2 minutes; positive pole in the other hand.) Successes which are said to have attended the suggestive treatment of warts, etc., have still to be verified, although such have frequently been reported.

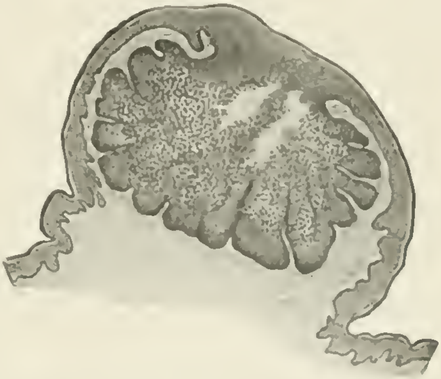
MOLLUSCA CONTAGIOSA (*Bateman*)

This affection consists in the appearance of flat raised nodules, whitish or slightly yellowish, on normal skin and covered with normal skin, of lentil to pinhead size; they are slightly indented in the centre and have a small punctiform opening from which a peculiar core can be expressed which consists of the so-called molluscum corpuscles. The affection, which is not rare in children is principally situated around the eye (eyelid), at the forehead, neck and genitals. It is to be regarded as a benign affection which is infectious and therefore transmissible.

Pathological Anatomy.—This is a new formation of epithelium of flabby consistency, the single lobules of which are separated by connective tissue cords: a proliferation of glandular epithelial ducts to the cutis. Neisser believes that the epithelioma has appeared in consequence of protozoa penetrating into the deeper layers of the rete: the parasite which Neisser believes to have found belongs to the sporozoa. Other authors look upon these cellular growths as degenerative manifestations of protoplasm.

Etiology.—Although opinions on the nature of the virus are not matured, there is no doubt after the successful inoculation experiments of Rezius, Vidal and Pick, that molluscum contagiosum is a contagious, transmissible disease. Recently M. Juliusberg has carried out successful transmissions upon the skin with the filtered fluid of the molluscum corpuscles. The contagious nature of this disease had previously been considered probable on account of its frequent occurrence in families and schools (Tommasoli 56 children in one asylum in Siena). In all these experiments the incubation period was very long.

Fig. 84.



Mollusca contagiosa. New formation of epithelium of flabby consistency, the indented depression and excretory duct at the top.

Diagnosis.—Flat warts and milium are principally to be considered. The diagnosis, however, is easy in consequence of the efflorescence being covered with normal skin, the depression in the centre and the small punctiform aperture. The **prognosis** is good. The **treatment** concerns itself with the evacuation of the small nodules by expressing and curetting them, application of galvanocautery and electrolysis. Where there are very many molusca, desquamation cures may be tried (resorcin paste 10 to 30 per cent., green soap).

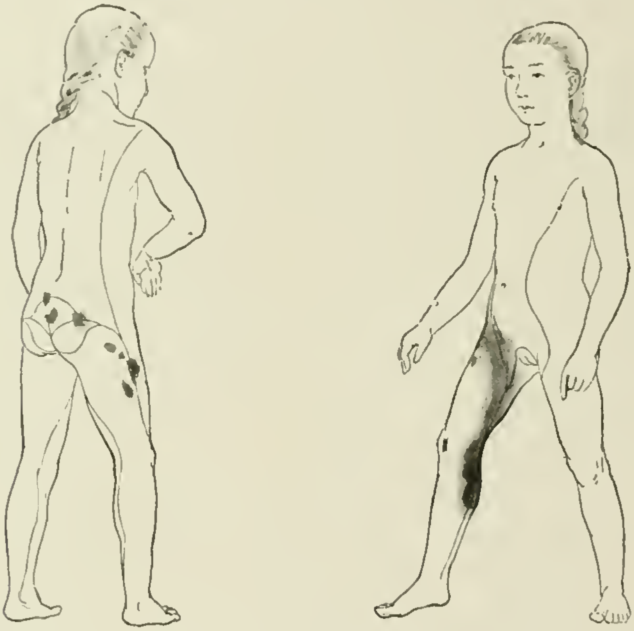
NÆVI

A nævus is a congenital new formation: that is, an affection which was either present at birth or developed later from a congenital disposition. According to their origin Neisser distinguishes between the true tissue nævi in which the neoplasm has originated by the proliferation of one particular portion of the tissue: and the organic nævi in which the neoplasm has been produced by the proliferation of certain organic parts of the skin. In nearly all tissue nævi there is more or less pronounced accumulation of pigment.

The simplest form of nævi are the *ephelides* (freckles), yellowish to dark brown spots, smooth and round or angular, from pinhead to lentil size, which appear on the exposed parts of the body, especially in the face and on the hands. Ephelides occur principally in summer, very often in spring when the sun shines somewhat strongly: and pale off in autumn and winter. Usually they do not occur before the fifth or sixth year, and frequently disappear spontaneously in later years.

In contradistinction to these growths the true soft nævi are to be described as prominent formations which by an increase of tissue are elevated above the superficial skin. We distinguish the simple nævus pigmentosus of black, brown or light color, the nævus pigmentosus verrucosus with a warty, knobby surface, the nævus pigmentosus spilus (with a smooth surface) and the nævus piliferus (covered with hair). The nævi grow with the body, and all forms may be found together on one individual. If a nævus extends over a large area of the body, we speak of a giant nævus; if it is besides covered with hair resembling animal skin, covering the abdomen, pelvis and femoral region, we speak of a bathing suit nævus (Kaposi). The smallest punctiform nævi are called lentigines.

Fig. 85.



Nævus linearis. (Systematized nævus) in the course of Voigt's demarcation lines.

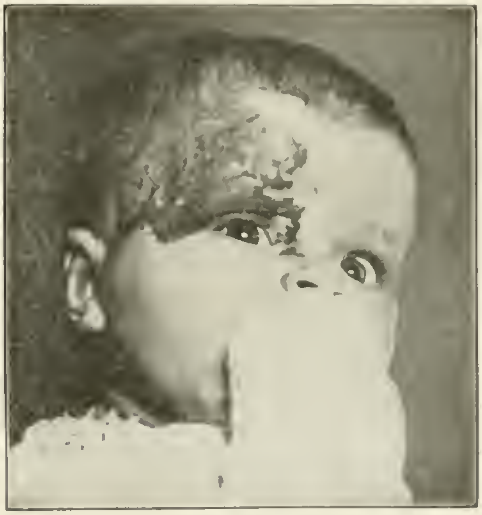
Besnier and Jadassohn have grouped a number of nævi which were described under the names of nerve nævi, nævus unius lateris (Bärensprung), nævus linearis verrucosus (Unna), together under the collective name of systematized nævi, in order to indicate that for the development of these nævi certain sites or conditions of development are influential. The systematized nævi are to be found in the course of nerves or of Voigt's demarcation lines (the branch regions of the skin nerves, Philipsohn), or in the course of the hair currents (following their lines of convergence and divergence), or in a certain metameric arrangement, especially at the back (Pecirea, Hallopeau), the arrangement being thus always different, but always demonstrable, and certainly influenced by the development of the skin. These nævi often produce considerable itching, they may also become easily œdematous and may during infancy cause trouble enough to render operative interference or galvanocaustic treatment necessary (Galewsky-Schlossmann).

Pathological Anatomy.—Histologically we find a large accumulation of cells in the cutis (nævus cells), a considerable accumulation of pigment in the rete and corium, and very often a strong proliferation of the cones and papillæ.

Etiology.—The cause of nævi is not known, although in many cases heredity can be demonstrated. In how far they may be occasioned by the pregnant mother receiving a psychic impression of similar growths, remains to be demonstrated.

Prognosis.—The prognosis of nævi in childhood is benign, although they sometimes require treatment on account of itching and development of eczema. Generally nævi develop gradually, then stand still, while in other cases they will gradually disappear. In advanced age they may be transformed into malignant ulcers.

Fig. 86.



Nævus flammeus.

Among the *organic nævi* those best known and most frequently occurring are the nævi vasculares (angiomatosi, flammei), the well-known circumscribed anomalies which originate from the vessels and are popularly known as fire moles. The simplest form of these are the punctiform or stellate telangiectases (nævi aranei) which are frequently found in the face of the youngest infants and consist of a hyperæmic central blood vessel and a number of smaller vessles which radiate from the same. The large fire moles may occupy large areas of the body and have a most disfiguring effect. The vascular nævi are either present at birth or develop later: often they disappear again after birth. They occur in all sizes and colors, singly or in groups, chiefly at the temples and cheeks, but also at the occiput, the root of the nose, the eyelids and the extremities, they are sometimes superficial and sometimes deeper, and may even spread to the mucous membrane. They may remain simple angiomata or form combinations with other tumors (sarcomata, etc.).

Among the glandular nævi in children there are the so-called nævi sebaceæ (adenomata sebacea), small tumors of the sebaceous glands in the nasolabial fold and at the cheeks, of red, yellowish or white color.

Concerning the **etiology** of angiomata we are likewise ignorant. Aside from the disfiguring effect and the very rare cases in which angiomata become malignant, the prognosis is good.

Treatment.—1. *Ephelides.*—The treatment of freckles demands protection against the rays of the sun and depigmentation. The protection from the sun in sensitive children (especially the blonde) is effected by wearing veils dyed with chrome or curcuma, or by painting the skin with a 10 per cent. quinine and gelanthum solution. Depigmentation is effected by corrosive sublimate, white precipitate, peroxide of hydrogen and oxychlorate of bismuth; for instance hydrog. peroxide 20.0 Gm., bism. oxychlori 0.5 Gm., sublim. 0.05 Gm., adeps lanæ 10.0 Gm., vaselin 20.0 Gm.

2. *Nævi.*—The treatment of nævi is one of destruction and can be carried out by cauterization with 1 to 10 per cent. sublimate collodium or trichloracetic acid or silver nitrate. The quickest effect is obtained by the Paquelin or galvanocautery. The best cosmetic results are obtained by electrolysis (positive pole in the hand, negative pole with the needle through the skin under the nævus, piercing in all directions two minutes each time, current of 5 M.A.).

3. *Angiomata.*—Their removal in the case of very small telangiectases in early infancy is best done, according to Unna, with ichthyol collodium (1 to 10 per cent., painting afresh as the cover is lifted). Larger angiomata are being excised, small and medium ones are removed by electrolysis (see above) or treated by the Finsen-Reyn lamp, or by radium exposures of 15 to 30 minutes duration (caution!).

DISEASES OF THE HAIR

APLASIA PILORUM INTERMITTENS (VIRCHOW)

(Monilethrix—Crocker. Spindle Hair)

This affection is distinctly hereditary and occurs in the early periods of childhood. It appears usually at the time when the hair changes, and falls out and only returns scantily. The skin becomes smooth, looks atrophic, and the scalp—almost bald—is covered with comedones and red acne papules which correspond with the follicles and are pierced by hair stubs. The hair is short, dry, glistening and shows light and dark tints, and the hair shafts become fibrous as in trichorrhexis.

Pathological Anatomy.—The hair shows fusiform swellings which are filled with air, alternating with strangulated sections devoid of medullar substance. The comedones are filled with doubled-up fusiform hairs (20 and more spindles in one comedo).

Etiology.—Bonnet has found a similar affection in horses. We only know at the present time that the affection is hereditary and has been observed in several generations (Lesser), and that usually children —almost exclusively male—inherit the disease from affected parents.

Treatment is useless in this chronic disease.

ALOPECIA AREATA (*Area Celsi-Pelade*)

In this affection, in which without assignable cause and without any noticeable change of the scalp, the hair falls out, circumscribed roundish foci make their appearance.

Usually the hair falls out first in a small annular disk, at the margin of which the hair breaks off, so that only small stubs and comedo-like bits remain in the sebaceous glands. The hair easily comes out when pulled, and the affection extends peripherally. New disks are formed which may become confluent and spread over the entire scalp. Gradually new unpigmented thin hair appears, which gradually acquires pigment and grows stronger. On the other hand the unpigmented hair may fall out again and again, until at last the final firm crop has been secured. In this way the affection may last for months before it is controlled. Aside from this benign form there is a malignant one in which within a few weeks the hair of the entire body may be lost.

Alopecia is found in children of all ages, but is comparatively rare (in Germany it is much rarer than in France). Sabourand distinguishes three forms in children: (1) In the first form the hair falls out slowly, reappearance also occurs slowly but only after five or six months. The affection commences with a spot, on which the skin looks atrophic, dry, somewhat desiccated (peladoide en aire unique avec atrophodermie excessive). (2) The second benign form seems to be contagious, occurs in families endemically without known cause; there are six to eight foci of the size of a quarter; reappearance of hair in six to seven weeks (peladoide familiale en petites aires multiples). (3) The true alopecia. It commences bilaterally at the occiput, spreads in secondary plaques over the head, is sharply symmetrical, lasts a considerable time, is cured spontaneously at puberty, persists sometimes for years and is not contagious.

Etiology.—Etiologically there are two opposite opinions, one favoring the nervous trophoneurotic and the other the parasitic theory, which is particularly upheld by Sabourand. For both views there are proofs and therefore alopecia probably has more causes than one (a nervous and a parasitic) which is also the opinion held by myself. Of interest for the etiology of alopecia are the experiments of Joseph (appearance of bald foci after cutting the second cervical nerve in cats) and the experiments of Giovanni, Buschke, etc. (appearance of alopecia after internal administration of thallium acet.).

The **prognosis** of alopecia areata is better in children than in adults, as the hair nearly always reappears on account of the absence of malignant conditions.

Treatment should be irritative and antiparasitic. The former effect is attained by massage and the faradic current. Among the chemical agents painting with balsam of Peru, tincture veratri, chinæ cantharides, capsici, argent. nitr. 1:15 with tincture of iodine or pure carbolic acid have an irritative effect.

The antiparasitic remedies, which should be applied together with the former, are the following: chrysarobin as a ½ to 10 per cent. ointment or dissolved in alcohol or chloroform, as a 30 per cent. ointment or its substitute, eurobin. A very good effect is attained by washing the head with sublimate, formalin or naphthol soap. The following treatment has given the best results in my hands: The scalp is washed daily with disinfecting soap and afterwards rubbed with an irritative ointment about 1 cm. beyond the margin of the affected place. The following ointment is serviceable. Chinæ cantharides, capsici aa 3.0 Gm. (45 gr.), balsam of Peru 0.5 Gm. (7½ gr.), ung. leniens ad 30 Gm. (1 oz.). Besides twice or three times a week in the evening the places are painted with chrysarobin or eurobin solutions, for instance eurobin 1.0 Gm. (15 gr.), acetone 30 Gm. (1 oz.), or inunction with the chrysarobin ointment stick, followed by the application of a cap to be kept in place over night.

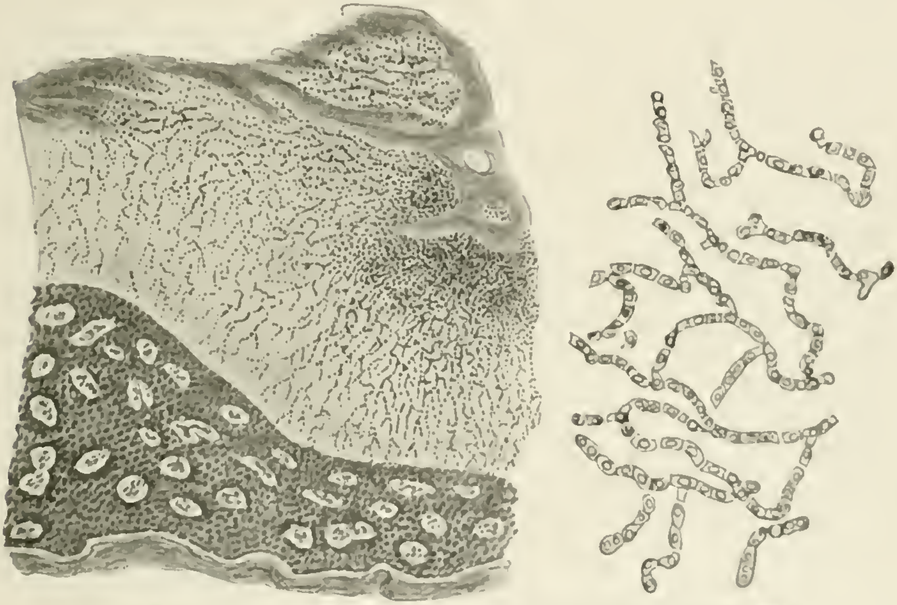
FAVUS

Favus is an affection caused by the achorion of Schönlein, in which small sulphur-yellow platelets are formed around the hair follicles. At first yellow punctiform growths appear on the scalp around the follicles which gradually increase in size to that of the yellow platelets, the upper surface of which is concave with a central depression while the lower is convex. These platelets, called scutula, always grow around the hairs, at first singly, afterwards communicating; they then lose their yellowish color and the scalp looks whitish gray, dry, dust-covered, and has an unpleasant odor, like the feces of mice. The skin under the scutulum is at first slightly hyperæmic, then becomes atrophic, whitish, and sinks in. The hair suffers likewise, loses its color, becomes thin and brittle, looks dusty, and falls out; the follicles and papillary bodies likewise become obliterated. The affection is chiefly situated at the head and in the hair of the head, but may spread over the entire body (often with a herpetic prodromal stage) and may even attack the finger nails which become brittle and splinter off. Favus is chiefly found in children of the poorer part of the populace in countries where cleanliness leaves much to be desired (Poland, Russia, Hungary). In Germany it practically only occurs when introduced from Poland; it is very rare in West and Central Germany.

Pathological Anatomy.—The scutulum is an accumulation of fungi in the corneal cells, the fungi being in the exterior root sheath of the hair. They invade the medullary space and in later stages lead to a pressure atrophy of the skin.

Etiology.—Favus is caused by the penetration of the achorion of Schönlein into the skin. The transmission takes place from person to person, but may also take place through mice suffering from mouse favus, and through their intermediate agents, cats.

Fig. 87



Favus scutulum with fungi; on the right fungi enlarged (Achorion of Schönlein).

Diagnosis.—The diagnosis of favus is easily established by the presence of the scutula and fungi. When the scutula are covered by grayish white scaly deposits, application of alcohol (Neisser) is sufficient to re-establish their yellow coloration. (Differential diagnosis with scaly eczema of the head.) Nor is the microscopic demonstration of the fungi in the scutula and hair at all difficult (unstained preparation made lighter by a 50 per cent. solution of potassium or stained preparations after Weigert).

Prognosis.—The prognosis of favus is unfavorable as soon as the fungi are situated in the hair, as it is not always possible to free the root sheaths of the hair from them or to remove the hair together with the root sheath.

Treatment.—The treatment consequently consists in removing the hair with the root sheaths, which is effected either by epilation with the forceps, by pulling out the entire hair by means of a pitch cap under chloroform anæsthesia, or by the application of the Röntgen ray. But as with the last-named treatment it is not always possible to secure the removal of the sheath, the X-ray will frequently prove ineffectual. The best method is the systematic epilation with the forceps. The operation completed, the head is bandaged with a 5 to 10 per cent. pyrogallus ointment and regularly washed with disinfectants, such as formalin or corrosive sublimate. Favus of the nails is treated with mercury plaster, hand baths, and painting with corrosive sublimate alcohol or pyrogallus acetone.

TRICHOPHYTIC DISEASES

Under the collective name of trichophytic diseases are embraced a number of affections which Sabouraud has the merit of having described separately and in detail. All the fungi seem to belong to one great family, and their exact description and differentiation should be studied in the text books. It may be mentioned, however, that Sabouraud divides the fungi into endothrix and ectothrix forms, according to whether the fungi or their spores penetrate into the hair or not; he further distinguishes them according to the size and form of the spores and the manner of spore formation (endospores, ectospores), according to whether such formation occurs within or at the end of the myeelia.

(*a*) GRUBY'S DISEASE

This affection, which is extremely rare in Germany and Austria, consists in the appearance of several round gray foci of 3 to 5 cm. diameter on the scalp, which are covered with gray scales. The hairs break off about 6 to 7 mm. from the root and are surrounded by a gray sheath like a cuff. The number of foci are from 2 to 10. This affection commences sometimes before the third year, rarely after the fourteenth year and has an average duration of eighteen months.

Etiology.—The microsporon Audouini (Gruby, 1843).

(*b*) TRICHOPHYTIA OF CHILDREN'S HEADS

This form of the affection begins with a prodromal stage which is very often overlooked. The skin in the vicinity of the scalp exhibits small spots of lentil size and vividly red color which gradually attain a diameter of 1 cm. They heal in the centre while the raised margin spreads peripherally. Often these spots are covered with small vesicles and scales. An examination of the scalp reveals the fact that the hair in a number of places is thinner, the skin at those places is scaly, diseased hair is mingled with the healthy, the broken stubs of the former boring themselves into the scales. Foci of this kind may exist to the extent of several hundred (each consisting of a few diseased hairs, sometimes only 3 or 10) or there may be a few larger foci where the diseased hair mingles with the healthy. The hairs are very difficult to pull out and can only be epilated singly. Their exterior is smooth; inside they are filled with spores of the trichophyton endothrix, breaking and splitting at the affected places. The affection extends beyond the period of puberty which has no influence upon it; its duration generally is eighteen months, but without treatment it may last two or three years or even longer. In Paris it is the most frequent form of trichophytic diseases.

Etiology.—The affection is caused by a form of trichophyton in which the spores are accumulated in the hair, resembling in their appearance a bag of nuts. The spores of trichophytia endothrix differ from those of the microsporon of Audouin in that they are larger.

Treatment.—The treatment of these two trichophytic affections is disproportionately difficult and absorbs months, because the hair is affected not only at the surface but also at the root involving the root sheaths, and because the hair can only with difficulty be epilated by reason of its breaking off easily in the attempt. To treat the complaint, the head should be shaved, after which strongly irritating remedies may effect the extirpation of the roots (tincture of iodine, oils and ointments containing croton oil). Besides ointments of chrysarobin and pyrogallus (1 to 10 per cent.), applied under a firm bandage, can be recommended. In how far treatment by the Röntgen ray will answer the expectations entertained in some quarters, is still uncertain; Sabouraud believes that the application of the X-ray may shorten the treatment by one-half.

(*c*) TRUE TRICHOPHYTIA

True trichophytia occurs but rarely in children, and then only in the second year of life. Its characteristic feature is the appearance of a specific inflammation of the skin, and not the destruction and annihilation of the hair, as was the case in the other two forms just described. The affection commences in a few well demarcated foci of vivid light red color; the plaques are ring-shaped, annular or serpiginous; they heal in the centre with light pigmentation or desquamation; and spread from the raised, circumvallate, light red margin. The circular plaques may have a concentral position, some of them may be confluent and be covered with vesicles. The affection which causes considerable itching is in children generally found at the head, but it may spread to the body and cause serious diseases of the nails. The treatment consists in epilation of the diseased hairs, should they be secondarily involved, and above all in the limitation and healing of the affected foci. This is effected by painting with tincture of benzoic sublimate (1 per cent.), painting with chrysarobin-traumaticin and by application of salicylic soap plaster or carbolized mercury plaster.

PITYRIASIS VERSICOLOR

Pityriasis versicolor is caused by the microsporon furfur described by Eichstedt in 1846. It is characterized by the formation of irregular yellowish to yellowish brown roundish spots which can be easily scratched off, exposing the skin which is only slightly reddened. The plaques may become confluent and occupy large areas. The face, palms and soles are always free, and the neck and dorsal surfaces of the hands are rarely attacked, the principal seat of the affection being on the trunk, back and arms. Its nature is benign; it is comparatively rare in children and they are never affected before the seventh or eighth year. It is hardly contagious, although its transmissibility has been established.

Pathological Anatomy.—On scratching off a scale and clearing it with a 30 to 50 per cent. solution of potassium, microscopic examination reveals in the horny masses the mycelia and gonidia of the microsporon furfur. The grape-formed accumulations of spores are characteristic.

Diagnosis.—The diagnosis is comparatively simple in consequence of the yellowish brown color, the typical seat at the chest and back and the microscopical findings.

Treatment.—The treatment of this benign affection consists in the application of baths, soaps and antiparasitic ointments. Patients should have ordinary or sulphur baths as frequently as possible, and be thoroughly rubbed with green soap or sulphurated naphthol soap, while in the bath. Once or twice daily the affected places should be bathed with 1 per cent. spirits of sublimate, 1 to 3 per cent. spirits of naphthol or 10 per cent. spirits of epicarin, which is followed in the evening by an inunction with sulphur naphthol ointment. As the spores also occur in the deeper corneal layers, relapses are a matter of course, if the treatment has only been superficial. To effect a complete cure therefore, a long-continued after-treatment with soaps (quinine, sulphur, sublimate soaps, etc.) is necessary.

ANIMAL PARASITES

SCABIES

The penetration of the scabies mite (acarus scabiei, sarcoptes hominis) is revealed by the formation of subcutaneous so-called scabies ducts, being fine small ducts 2 to 15 mm. in length and of whitish color. The skin is either of normal appearance or raised in the shape of a vesicle or hyperæmic papula. It may assume a dark appearance from dust and dirt and in consequence of the intense itching there are scratch marks, signs of inflammation of the skin, and small pus pustules to be seen everywhere between the ducts. The affection is chiefly situated between the fingers, around the wrists, in the axillary folds, at the nipple, navel, and penis, at all places subject to pressure (belt, etc.); in little children also at the plantar surface and the interior edge of the foot. The face is always free. The affection which in the beginning is difficult to recognize, spreads comparatively rapidly, and there are complications such as furuncles, impetiginous eczema, glandular enlargements, etc. The disease is highly contagious and children are principally infected by sleeping with adults who suffer from scabies. Consequently it is met with in every stage of childhood.

Anatomy.—The mite which is about 0.2 to 0.3 mm. in length, can still be seen with the naked eye, burrows itself into the skin and forms the so-called scabies ducts where the female deposits its eggs. The male lives in small fossæ in the vicinity of the ducts. The latter contain the oval eggs and brownish granules—the feces of the mites.

Etiology.—The disease is spread by direct transmission (cohabitation). In how far clothes and body linen may transmit the disease has not yet been established. Alexander has also found children suffering from dog scabies, which consist of a papulovesicular exanthema, and lasts six to eight weeks, healing spontaneously and easily controlled. There are no typical ducts, and mites are not always demonstrable.

Diagnosis.—The diagnosis is secured by the demonstration of the ducts or the mites. The skin is smeared with ink to make the ducts more visible and the duct is pricked with a needle, upon which the mite will be found at the blind end of the duct. If a microscopic examination of the duct is desired, it is removed with a pair of scissors and examined in a 10 per cent. solution of potassium.

Prognosis.—In children the prognosis is favorable, but often in consequence of the strong antiparasitic ointments a dermatitis resembling scabies may remain and continue to exacerbate if the treatment for scabies is persisted in.

Treatment.—Among the available remedies balsam of Peru and its derivatives stand in the first place. For children inunction of the body with balsam of Peru 20 Gm. (5 dr.) olive oil 100 Gm. (3 oz.) on several successive evenings is advisable. If there is considerable dermatitis, the ointment should consist of ung. zinci 50 Gm. (1½ oz.), balsam of Peru 2.0 to 4.0 (30–60 gr.). Substitutes emitting less odor are peruol and peruol soap which should be applied daily. Styrax is similarly applied (10:20 Gm. olive oil), while the strongest remedy consists of naphthol ointment (Kaposi), axungia porci 100 Gm. (3 oz.), sapo virid. 50 Gm. (1½ oz.), naphthol 15 Gm. (½ oz.), sulf. præcip. 10 Gm. (2½ dr.) d.s., to be used as an inunction every evening. In the place of naphthol a 10 per cent. epicarin ointment may be used. After 3 or 4 days' application of these ointments the children are given a bath, and as after-treatment to heal the dermatitis, 5 per cent. salol oil (with castor oil 40 per cent.) or zinc ointment (zinc oil or paste) is applied. As a matter of course, careful attention should be paid to changing and disinfecting the clothes and body linen, and in the case of a family epidemic the entire family should be treated simultaneously.

CREEPING ERUPTION (*Lee*). CREEPING DISEASE (*Crocker*)

The creeping eruption consists in the appearance of slightly raised, light red, very long ducts under the skin, into which the larvæ of an insect (probably gastrophilus) bore themselves mole-fashion, and creep forward. The affection is very rare and outside of Russia it has only been described in children (Rille).

Treatment.—Excision, electrolysis.

PEDICULI

(*a*) PEDICULI CAPITIS (HEAD LICE)

The pediculi and their whitish gray nits are located on the hairs of the head, producing intense itching and irritation. They may easily be accompanied by an eczematous condition of the scalp described as plica polonia (see eczema, page 468). The treatment of simple pediculosis consists in washing the head with petroleum or 5 per cent. naphtha. Acetic sublimate (1:300), acetum sabadillæ (which often produces intense burning) may be applied in the evening under a bandage. In the morning the scalp is washed, and the entire procedure repeated for several days. (For treatment of simultaneous eczema see p. 475.) The hair should not be cut off except in case of need; the nits are removed by combing with a narrow toothed comb and washing the hair with vinegar, which will losen the nits.

(*b*) PEDICULI PUBIS (MORPIONES)

Pediculi pubis occur very rarely in children. Trouessart, Heisler, Grindon and others have found morpiones in the eyebrows of children, the cilia and the scalp. The children's age varied between fourteen months and twelve years, Grindon describing an entire family epidemic of this description. Should the parasites be located in the cilia, they may produce considerable blepharitis, hyperæmia of the eyelids, etc. In one of these cases up to 100 insects were found on one eyelid (Jullien).

Treatment.—The treatment of morpiones on the scalp is effected with acetic sublimate, acetum sabadillæ or ung. hydrargyri album, those of the eyebrows with yellow mercury paste.

(*c*) PEDICULI VESTIMENTORUM

These lice have their favorite seat in the folds of the body linen and only invade the skin in order to feed. They are met with in places where the clothes are in close contact with the body, at the waist (sacrum), also in neglected uncared for children. After a time the skin becomes pigmented owing to scratching.

Treatment.—Cleanliness, baths (sulphur baths), disinfection of clothes and linen in dry heat 70° to 80° C. (138° F. to 177° F.) and the use of insect powder.

Pulex irritans produces the punctiform hæmorrhages on the skin which are known as fleabites. On sensitive skins they may cause slight wheals.

The *bed bug* (cimex lectuarius) causes papulous manifestations of the skin. To destroy both parasites use insect powder and wash with spirits of menthol and thymol.

To treat the stings of gnats (culex pipiens) and crane-flies apply sal-ammoniac, ammonia, ichthyol collodium, while as a prophylactic application a solution of chysanthemum seed in alcohol can be recommended.

TUBERCULOUS DISEASES OF THE SKIN

BY
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INTRODUCTION

SINCE the discovery of the tubercule bacillus by Koch, we have come to recognize many forms of skin tuberculosis, in addition to the acute ulcerative forms. Lupus, scrofuloderma and tuberculosis cutis verrucosa are the most important of these.

The characteristic histological picture of the miliary tubercle with giant cells and epithelial cells with a tendency towards caseation, the positive bacteriological findings, and the experiments on animals, have done away with all doubts as to the tuberculous origin of these diseases.

Besides those lesions which are genuinely tuberculous in character there are still left many conditions which may be observed clinically in individuals supposed to be tuberculous. The nomenclature of these has always caused the greatest difficulty. They are partly grouped, however, under the heading *acne scrofulosorum.*

In 1891, Barthelemy proposed the name folliculitis or acnitis for these particular cases, on account of their supposed origin in the follicles; but he did not strictly raise the question of their relation to tuberculosis. To Darier is due the credit of having called particular attention to the tuberculous origin of these lesions. He combined these dermatoses under the name *tuberculide.*

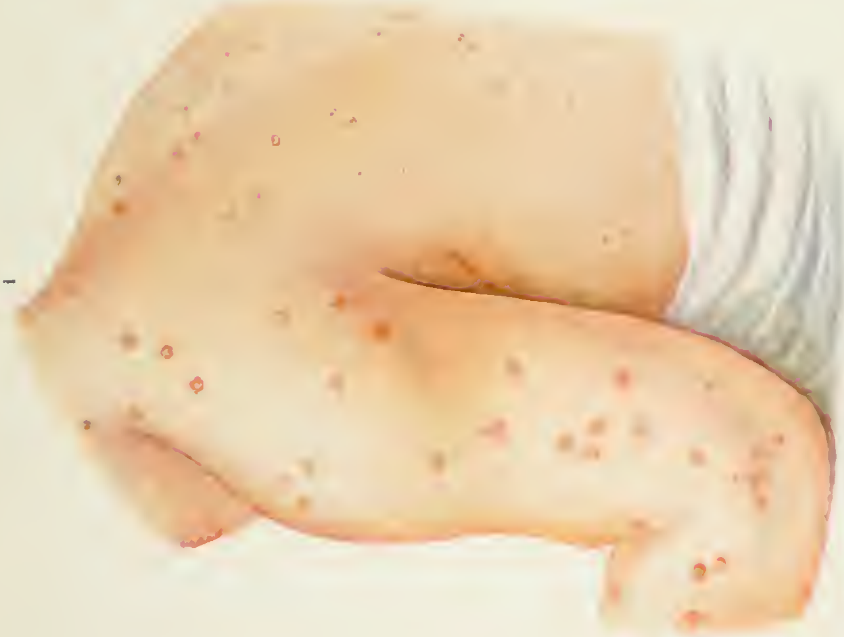
Now commenced a period of zealous study of these diseases and their causes. Long treatises were written from this theme and these gave occasion for many sharp discussions and controversies at the Congress at Paris in 1900. At that time Boeck proposed a division of these conditions into perifollicular, superficial, and nonperifollicular deep rooted tuberculide, and combined the whole group under the designation of the exanthemata of tuberculosis.

However, as a united action in regard to the nomenclature was not reached, I will add another name proposed by Pautrier, *i.e.* "*Les tuberculoses cutanées atypiques.*"

Thus it may be noted that the clinical conception of these dermatoses after the basic labors of Darier, Boeck, Hallopeau and others, was generally accepted.

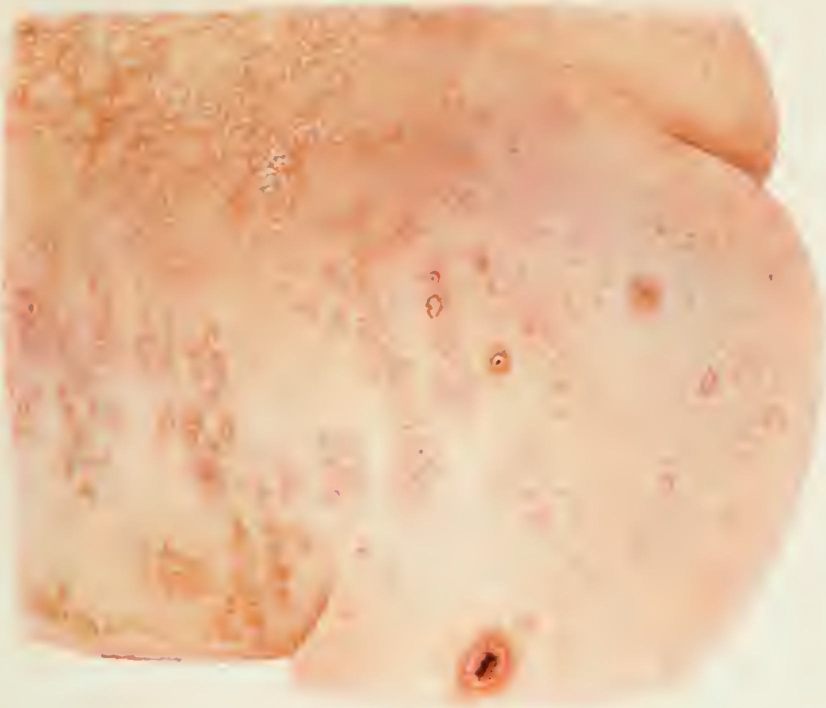
PLATE 68.

I



Papulo-necrotic tuberculid

II



Lichen scrophulosorum

To the above-mentioned conditions may be added the long well-known picture of the *papillary necrotic tuberculide.*

The chief characteristic of these forms is comprehensively expressed by Zollikofer, as a tendency to gradually spread out over the territory of predilection; a chronic condition resulting; which may run along for weeks, showing apparently but slight inflammatory reaction all the time.

Another peculiarity exists, in that these lesions remain inoffensive. That is to say, they are without contageousness towards healthy tissue. They show no disposition to invade neighboring healthy districts, either upon the surface or deeper.

While the combination of these dermatoses with tuberculous diseased organs is acknowledged by almost all authors, chiefly on account of their clinical observations, and that these lesions occur only in those who are tuberculous; still the question of their origin forms the subject of a lively controversy to-day.

In the beginning, when the disease was first recognized and the name tuberculide was just being evolved, Hallopeau accepted the toxine hypothesis and tried to draw a sharp contrast between these forms and the old bacillary skin tuberculosis, by coining the word toxituberculide. According to him, the infective material (analagous to bromine or iodine poisonings) coming from distant tuberculous foci lying upon the skin, would travel by the circulation in the epidermis, and being deposited would cause atypical changes which were called tuberculide.

The apparent symmetry with which these eruptions appear on both sides of the body, caused Boeck to form the theory of *Angio-Neuroses.* He did this by placing the attacking points of the toxin, not directly in the blood vessels, but in the vasomotor nerve centres, an opinion, however, which Boeck himself dropped later on.

Jadassohn opposed the toxin theory, relying chiefly upon the findings of Philippson in cases of tuberculous thrombo-phlebitis. Independently of him, Darier put himself on record for the bacillary origin of the tuberculide. Indeed he goes so far as to express a belief that it is the bacilli which are weakened in their virulence or even dead bacilli which when deposited in the skin will lead to the various forms of the tuberculide.

The ever increasing frequency of the finding of bacilli in lichen scrofulosorum and the other forms of tuberculosis, is the cause for the acceptance of a bacillary origin for these dermatoses; and in fact, this is now very generally believed (Jadassohn, Neisser, Comby, Nobel).

On the principle of these hypotheses, partly formed by histological and bacteriological findings, and partly by positive inoculative experiments; we cannot speak any more of a sharp contrast between the old skin tuberculosis and the so-called tuberculide, but must trace back, particularly the homogeneous forms, from the disseminated miliary exanthem with plentiful bacilli, to the lichen scrofulosorum; from the multiple post-exanthematous lupus to the erythema induratum—all of these to a blood infection with bacilli in the most various grades of virulence and vitality.

The difficulty of making a very early diagnosis of tuberculosis of the apices of the lungs; or of the so frequently occurring tuberculosis of the bronchial glands in childhood, is well known. By a most minute examination of the skin we are at times rewarded by the finding of isolated or multiple foci of lichen scrofulosorum, follicular efflorescences or disseminated lupus. The finding of any one or more of these would show us the existence of a latent tuberculosis and thus put us on our guard and suggest the therapeutic remedies which used thus early, would save life by relieving the patient of the danger threatened by a new exacerbation or general spreading of the tuberculous process.

The most frequent forms of skin tuberculosis occurring in childhood, are lupus vulgaris, and scrofuloderma.

LUPUS

The characteristic sign of lupus is the so-called lupus nodule which is made up of a small circumscribed group of miliary tuberculous nodules and generally has its seat in the capillary layer of the skin and at times going deeper, into the corium.

Clinically, it presents a flat, brown infiltration. The brown color may be more distinctly brought out by the pressure of a cover-glass.

Pressure occurs, followed by infiltrations, by the confluence of neighboring primary lupus macules which are mostly round and disk-like; these cause frequent interruptions of the nutrition of the epithelium and of the exfoliations. On the edges of these disks, new eruptions of nodules often occur, frequently as *lupus serpigineous.*

When both the capillary layer of the skin and the epithelium participate in the whole process in a marked degree, we have resulting those papillary tumors called *lupus verrucosus papillomatosus.*

Besides the above-mentioned retrogressive epidermal metamorphoses, it sometimes happens that deeper changes take place and we have a softening and decay of the nodules and the formation of the characteristic lupus ulcers.* The surfaces of these ulcers are generally covered with a thick yellow scab. If much discolored by blood the scab is of a darker hue. After the removal of the scab, the uneven floor of the ulcer is seen, which generally bleeds easily. The border of the ulcer is hemmed in by the miliary lupus nodules.

The mucous membranes may be the seat of the primary areas. In this location it generally forms diffuse infiltrations. The mucous membrane appears gray, uneven and granulated and ulcerations or deep fissures develop. Extensive destruction and mutilation of the tissues may occur during the advancement of the lupus processes in the mucous membranes as well as in the skin.

---

* This is called *lupus ulcerosus*

Location.—Generally in childhood, the lupus appears in isolated areas which show according to their location a corresponding favorite form. For instance, the miliary and maculous forms have a predilection for the face; the lupus verrucosa for the tissues about the joints, etc.

Again, very frequently in childhood, the disease is found scattered here and there over the entire body. This is particularly frequent after the acute exanthemata such as scarlet fever or measles or chicken-pox. This has been named *lupus vulgaris post exanthematicus.*

While most authors accept the theory of the endogenous hæmatogenous infection for all these forms of disease, Unna and his followers remain true to the old accepted theory of inoculation tuberculosis, even for disseminated lupus.

Cases have been reported from various sources, which like the rest of the tuberculous skin diseases, when seen in connection with the acute exanthemata or occurring afterwards, have to be studied under children's diseases. Lately Tobler has compiled all of these cases which are mentioned in the literature.

I also have had occasion to observe two cases of disseminated lupus following attacks of measles. The first case showed more than fifty lupus nodules (foci); which were spread here and there over the whole body. A periproctitic abscess was also present. In the second case the foci were less in number but the child showed several maculous areas on the face, and isolated areas on the back of the hands, which were somewhat different in appearance from the ordinary form of lupus and which corresponded more closely to tuberculosis cutis verrucosa.

Tuberculosis cutis verrucosa is observed in patches, which are partly covered with papillary dirty brown or gray colored excrescences and partly with small pustules or scabs. Anatomically a circumscribed or diffused infiltration is observed, preponderating in the capillary layer of the skin. Epithelial cells, giant cells and some caseation are also observed. In this condition, tubercle bacilli are ordinarily found with ease and in large quantities; quite in contrast to lupus.

The **differential diagnosis** between the serpiginous form of syphilis and this condition presents some difficulties; the more so, as the serpiginous form of syphilitic lesions may be present for months or even years and healing may have occurred, leaving only fine epidermal scars as a result. A careful examination of the generally somewhat deep scars, sometimes discloses typical lupus nodules, and the finding of these make the diagnosis sure.

Sometimes the use of the tuberculin injection, as recommended by Neisser, is followed by good diagnostic results. With the disseminated forms, one often observes at the first moment, a more or less remote similarity to acute universal psoriasis of the capillary layer of the skin, which is so often seen in childhood. The absence of the small scales, after the removal of which, in psoriasis the capillary layer of the skin easily bleeds; and the color of the single efflorescence, often also the various typical patches of psoriasis observed here and there over the surface of the body, are quickly decisive for purposes of diagnosis when carefully considered.

SCROFULODERMA

According to Lang the relative frequency of the occurrence of scrofuloderma and lupus, is in doubt. The origin of scrofuloderma is ordinarily in the superficial fascia and its characteristic form is the scrofulous gummata or scrofulotuberculide. Even in early infancy, we find, particularly in the skin of the face, bluish red or brownish red nodules which may be seen more or less projected over the surface of the skin. These are softened in the centre and are not painful to the touch. The nodule lying subcutaneously, develops at times from a gland and raises slowly towards the surface of the skin which then becomes adherent to the subcutaneous infiltration. Sometimes after months of torpid existence, a subinvolution of the infiltration without perforation slowly develops. More frequently an eruption through the very much thinned skin takes place and an ulcer is formed. The floor of the ulcer is filled with easily bleeding, yellowish gray granulations or with necrotic-like tissue. This is called *scrofuloderma ulcerosa* (Plates 60–61).

The process of healing follows slowly with cicatrization. The scars are thin, delicate, and white; sometimes bluish on the edges and frequently partially covered with a scale and healing in an irregular manner.

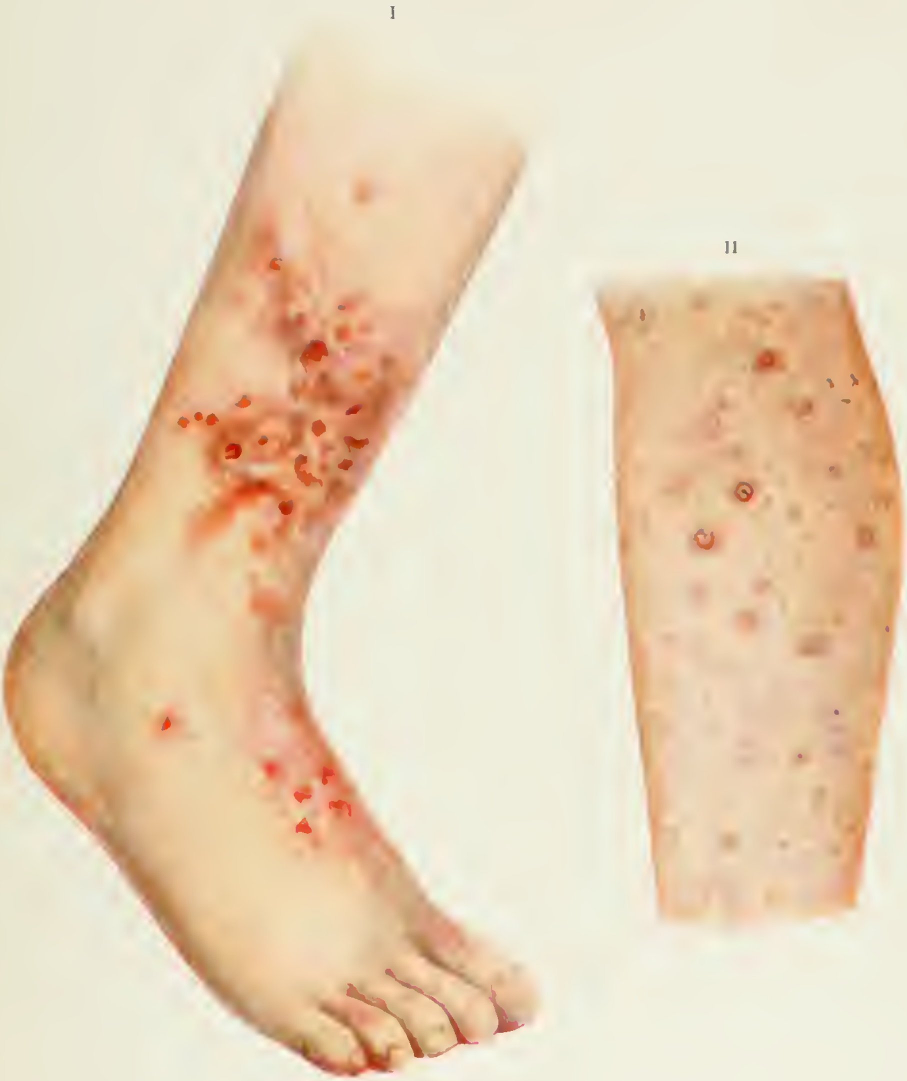
Besides appearing in very isolated areas, often over the seat of tuberculous glands in the neck, it may develop by spreading from a gumma or from a primary lupus area, along the lymphatic vessels and so we have a new formation of scrofuloderma.

In a few cases we observe scrofulodermata spread over the whole body; which makes probable a hæmatogenous invasion. We have observed this in a case after scarlet fever.

In its **anatomical relations,** scrofuloderma shows a sharply limited tuberculous infiltration, containing a great number of giant cells and generally only a few tubercle bacilli.

In making the **differential diagnosis** it is sometimes difficult to distinguish between this disease and erythema induratum (Bazin); which also makes its appearance in subcutaneous nodules; but this latter disease is localized in the lower extremities, and in contrast to scrofuloderma, it very seldom reaches the stage of perforation. Besides, the erythema induratum very rarely occurs before puberty.

PLATE 69.



I. Scrophuloderma ulcerosum.
II. Folliculitis after measles.

LICHEN SCROFULOSORUM

I now desire to mention some forms of tuberculosis of the skin which frequently occur in childhood and which formerly were separated from the original forms of tuberculosis on account of the negative findings of bacilli and which were counted as belonging to the closer group of the tuberculides.

Lichen scrofulosorum is characterized by groups of follicles, appearing as nodules of a pale yellow or yellowish brown color; which project only a little above the surface of the skin; and which are often covered with fine scabs. These are found particularly upon the trunk, sometimes upon the extremities and in one of my cases, upon the scalp.

This disease is found in young individuals in about ninety per cent. of all cases suffering with scrofulosis, tumors of the glands or other forms of scrofulous tuberculosis, according to Hebra.

Histological examinations, and also positive animal inoculations, have proved the tuberculous nature of the disease (Jacobi, Wolf, Pellizari, Bettmann).

The view originally held, was that this exanthem represented nothing more than the expression of a general cachexia.

Lichen scrofulosorum, like the rest of the tuberculides, appears after the acute exanthema or in conjunction with exacerbated tuberculosis. In this connection, we saw a boy who developed a lichen scrofulosorum, which spread over the whole body, even over the scalp, and at the same time showed an exacerbation of a spondylitis. The tuberculin injection (0.0001 old tuberculin) resulted in a slight rise in temperature, and also in a local reaction of the lichen area which appeared larger and more vividly red.

Further, we observed a case in conjunction with measles, in a child two and a half years of age. There appeared large grouped areas of lichen scrofulosorum on the trunk and extremities, three weeks after measles, for which the child had been treated in our hospital. Besides this, the child showed numerous papillary necrotic nodules on the extremities, follicles, swelling of the glands of the neck, conjunctivitis, eczema and rhinitis. The general health was very bad for weeks, the more so as there was besides a hæmorrhagic diathesis forming dot and splash-like skin hæmorrhages. The evening temperature ranged about 39° C. (103° F.) It was interesting to observe that in the follicular nodules in which the bleeding occurred, the necrosis made deeper progress and ulceration developed. (See Plate 69. Lichen scrofulosorum folliculitis with ulceration and purpura).

After lasting for months the process slowly healed; the folliculitis leaving shallow scars. The tuberculin injection (0.0001 old tuberculin) which was given before the child left the hospital, resulted in a rise of temperature and a slight deterioration of the general health.

The combination of lichen scrofulosorum on the trunk with papillary necrotic tuberculide on the extremities which was well demonstrated in the foregoing case occurs quite often.

Where the nodule is found in a subcutaneous or intercutaneous position the extremities are found to be covered to a more or less degree, with an exanthem, whose primary efflorescence represents inflammation of the nodule. Gradually these nodules, which are of a bluish red or yellowish red color, rise from the deeper layers of the skin and show small indentations on their surface, which are covered either with white scabs or small crusts. The nodules remain for months, slowly flattening, then gradually become paler in color and at last disappear, leaving behind a somewhat depressed, flat scar; which seems to be surrounded by a slightly raised peripheral wall. Folliculitis, therefore, may be said to consist of an inflammation resulting in necrosis, scars and some atrophy. This condition is generally localized; mostly on the extremities, over muscles of extension, on the outer surfaces of the arms, on the backs of the hands and fingers, on the upper surfaces of the feet, on the backs of the ankles and legs and on the ears.

It develops in many children suffering from tuberculosis, first appearing on one or the other favorite locality, as an isolated tuberculide. We often notice these areas of infection on the extremities of children brought into the hospital, suffering from tuberculous meningitis. In the course of meningitis due to other causes, we have never noticed these follicular eruptions.

Sometimes, the development of skin tuberculosis does not occur probably because the duration of the tuberculous disease is too short, and the condition of the skin for the sowing out of the eruption, too unfavorable.

At times a disseminated eruption covering the whole body occurs, particularly after infectious diseases. Measles may be mentioned as the chief of these. On the fourteenth day of July, 1905, a child sixteen months old was presented for hospital treatment. Two weeks before the child had measles and was running a somewhat chronic fever. It was now attacked by an exanthem, which proved to be a typical papular necrotic universal tuberculide, following the measles. In the further course of the disease in this case, several quite typical follicular nodules were observed, particularly in the face. These became much inflamed and softened in the centre from secondary infection with staphylococci. An injection of 0.0001 old tuberculin resulted in a slight rise of temperature; but did not produce local reaction.

Almost all authors agree that this form of tuberculide is not of toxic but of bacillary origin. It may be that the bacilli are present in very small numbers; or that they possess a weakened virulence, or they may be in a dying condition (Darier, Jadassohn, Zollikofer).

In the **differential diagnosis**, acute acne vulgaris has a certain similarity in appearance but differs from it as follows: acne vulgaris is always adherent to the follicles, that is to say, causes a typical pustular formation, and in healing seldom leaves a scar.

When localized around the mouth or anus, miliary ulcerative skin tuberculosis must also be excluded; this form being often found in these situations, as well as in other locations, it is generally combined with tuberculosis of the lungs or intestines. This condition is characterized by a sharp-edged, small, shallow ulcer, quite in contrast to folliculitis. These ulcers contain numerous tubercule bacilli.

PROGNOSIS AND TREATMENT OF TUBERCULOUS DISEASES OF THE SKIN

It is advisable in discussing the prognosis of tuberculous diseases of the skin, to use the old division, separating the older known forms of skin tuberculosis from the tuberculide. While the prognosis of the former, particularly in case of lupus, is more or less unfavorable, for if left to itself, it may cause the most severe and irreparable destruction of tissue; in the latter, the tuberculide, the prognosis is much more benign. Here even the disseminated forms may eventually result in spontaneous healing. The disease, spreading in patches or in deep ulcers, has never been observed. Therefore, while these last forms are not very dangerous to the individuals affected, they are of considerable importance in regard to prognosis, for they point to the existence of tuberculous foci and stamp the patient as tuberculous.

As regards the treatment of these tuberculides, lupus occupies an exceptional position, no matter whether only isolated nodules are present or only a few disseminated areas of the disease, we have only one proper therapeutic measure and that is the speedy removal of the lupus areas.

The best method is early excision, which may be employed with success, even when numerous areas are present. In one of my cases more than fifty were removed. If in the removal of the larger diseased areas there should occur considerable loss of tissue; skin grafting after Thiersch's method may be done. Krause uses long incisions, thus making pedunculated flaps, easily stretched.

The Vienna school (Lang) particularly deserves great praise for the elaboration of these methods.

The other therapeutic measures, while they occupy a secondary place, nevertheless are followed by good results. They consume too much valuable time. Among these may be mentioned the Rontgen rays and Finsen's light therapy treatments. Sometimes the tuberculin injection which Neisser has used with success gives good service in aiding us to recognize the lupus nodules.

Surgical treatment is also the best for cases of scrofuloderma.

Here we get the desired results by enucleation done with a sharp spoon, the resulting wounds being covered with salves. Boro-vaselin, dermatol or iodoform vaselin may be used.

In this connection it would be wise to mention, that in rare cases an outbreak of miliary tuberculosis is observed after operative procedure.

In the remaining forms of skin tuberculosis, general treatment holds the foremost place and must be constantly borne in mind.

With all these remedies at our disposal, we must strive for an improvement in nutrition. We often notice that after a short stay in the hospital, under suitable care of the skin and proper nourishment, tuberculides heal quickly and the child gains in weight. In other cases of a more torpid character, we only see good therapeutic results after a long course of treatment with salt and iodine baths, after giving creosote and codliver oil preparations and after prolonged stay in the country or at the sea-side.

As a matter of course, the healing of any particular tuberculide is not always identical with the healing of the primary tuberculous organ and after a shorter or longer interval a fresh outbreak of the skin tuberculosis may occur.

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